Journal of Pediatrics and Neonatology

ISSN 2688-5573

Case Report

Double Aortic Arch in Developing Countries: Diagnostic Delay and Clinical Lessons

Mohameth Mbodj[1], Idrissa Demba Ba[1,2], Serigne Tawa Ndiaye[1], Amadou Sow[1,3], Mohamed Fahd Negra[1] and Ibrahima Diagne[4,5]*

[1]Albert Royer National Children's Hospital, Dakar, Senegal

[2]Cheikh Anta DIOP University, Dakar, SENEGAL

[3]Centre Hospitalier Abass Ndao, Dakar, Senegal

[4]Gaston Berger University, Saint Louis, Senegal

[5]Hôpital Régional de Saint Louis, Saint Louis, Senegal

Abstract

Double aortic arch is a rare congenital malformation. It is the most common cause of symptomatic complete vascular ring. Its signs are non-specific, making diagnosis difficult.

We retrospectively analysed 3 files of patients hospitalised at the Albert Royer Hospital for double aortic arch during the year 2020. We collected data concerning the circumstances of discovery, the time to diagnosis, the clinical signs and the therapeutic modalities. The mean age at diagnosis was 8 months. Clinical signs at the time of diagnosis were dominated by respiratory distress and stridor. The interrogation revealed recurrent respiratory infections. All 3 patients underwent surgical management.

Introduction

Double aortic arch represents less than 0.1% of aortic arch variants. It arises from the lack of involution of the dorsal caudal aorta [1]. Aortic arch anomalies are rare and account for 1% of cardiovascular congenital anomalies [2]. The extent of symptoms depends on the space between the two aortic arches. The onset of symptoms is usually early, before the age of 3 years, and dominated by non-specific, respiratory and gastrointestinal signs and the extent of symptoms depends on the space between the two aortic arches [2,3]. Imaging plays a major role in the diagnosis of double aortic arches, as well as in determining the type of aortic arch and its relationship to respiratory and oesophageal structures, and associated abnormalities [4]. There is little data on aortic arch anomalies in Africa. We report on the difficulties in diagnosing double aortic arches in three cases.

Observations

Case 1

A 15-month-old female infant with good psychomotor development from a 42-year-old mother with 1 abortion, pregnancy poorly monitored, delivery at term, Apgar 9/10, no parental consanguinity, admitted for persistent respiratory difficulties and coughing during feeds. She had a history of hospitalization at 2 weeks of life for respiratory difficulty, and 3 recurrent episodes of bronchiolitis. There were two sibling deaths during the neonatal period of unknown cause. On admission she had a weight of 8.9 kg, height 75 cm, fever 38.5°C, tachycardia 150 beats/min, polypnoea 50 cycles/min, hypoxia 85% on room air, signs of respiratory struggle: nasal flaring, intercostal draught, stridor. Pulmonary auscultation revealed basal right crepitus and diffuse sibilant rales in both lung fields. Biological tests for BAAR and GenXpert in the gastric fluid were negative. There was a hyperleukocytosis of 23400 with a predominance of PNN and a positive CRP. Cytobacteriological examination (ECB) of the secretions + culture had found a non-groupable Streptococcus.

On chest X-ray, there was a basal alveolar-interstitial opacity on the right with blunting of the homolateral cul de sac and horizontalization of the ribs on the left.

Laryngotracheobronchoscopy revealed diffuse inflammation of the laryngotracheobronchial mucosa and purulent, viscous tracheobronchial secretions. The diagnosis of superinfected asthma associated with bacterial laryngotracheobronchitis was retained. The persistence of respiratory distress after 10 days of treatment prompted a thoracic CT scan to look for an associated bronchopulmonary malformation (Figure 1).

The thoracic CT scan showed a double aortic arch with a predominantly right arch, each arch giving rise to a subclavian and a primary carotid artery, a mass effect on the thoracic oesophagus in contact.

Cardiac ultrasound revealed a double aortic arch image with no other cardiac abnormalities.

The infant was operated on at the age of 2 years. The postoperative course was straightforward with no complications.

Case 2

A 2-month-old female infant followed in the department for laryngomalacia was admitted to hospital with fever and respiratory distress. She was born to a 34-year-old mother, 3 gestations, 3 pares,

Citation: Mbodj M, Demba Ba I, Ndiaye ST, Sow A, Negra MF, Diagne I. Double Aortic Arch in Developing Countries: Diagnostic Delay and Clinical Lessons. J Pediatr Neonatol. 2025;6(1):1051.

Publisher Name: Medtext Publications LLC

Manuscript compiled: Jul 22nd, 2025

***Corresponding author:** Mohameth MBODJ, Albert Royer National Children's Hospital, Dakar, Senegal, Tel: 00221 77 187 26 01

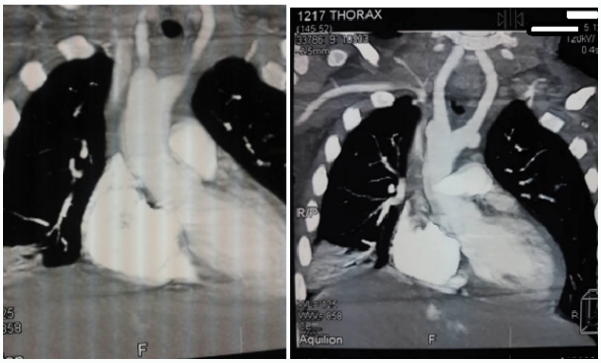

Figure 1: Chest CT scan showing a double aortic arch with a predominantly right arch.

gravidic hypertension. Pregnancy followed with 3 ANC carried to term. The newborn was admitted to hospital at the age of 20 days for management of acute laryngitis on a background of laryngomalacia slightly improved with oxygen therapy and adrenaline spray. On admission, she presented with weight = 6 kg, infectious syndrome with fever 39.1°C, tachycardia 150 beats/min, RF = 42 cycles/min, hypoxia 92% on room air, mild respiratory distress, stridor, right basal lung condensation syndrome. On biology, there was a predominantly neutrophilic hyperleukocytosis of 20,700 with a positive CRP. The chest X-ray showed a right basal parenchymal opacity.

The diagnosis of pneumonia in a laryngomalacia setting was made.

The infant was started on dual antibiotics (cefotaxime/amikacin), oxygen therapy and nebulised adrenaline spray.

The evolution was marked by a regression of the infectious signs and the persistence of a light respiratory distress and stridor motivating the realization of other explorations within the framework of the etiological research.

Laryngoscopy revealed a bulge in the left wall of the trachea, resulting in extrinsic tracheal stenosis.

The oesogastroduodenal transit (TOGD) showed a left posterolateral impression of the thoracic oesophagus at D3 in favour of an abnormal vascular arc (Figure 2).

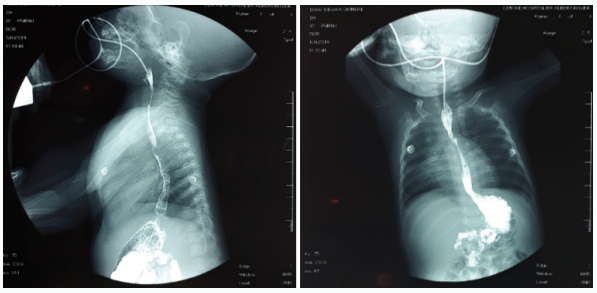

Figure 2: Oesogastroduodenal transit showing a left posterolateral impression of the thoracic oesophagus.

Thoracic CT angiography showed a non-functioning double aortic arch with a retro-oesophageal diverticulum, probably due to compression of the aerodigestive arch (Figure 3).

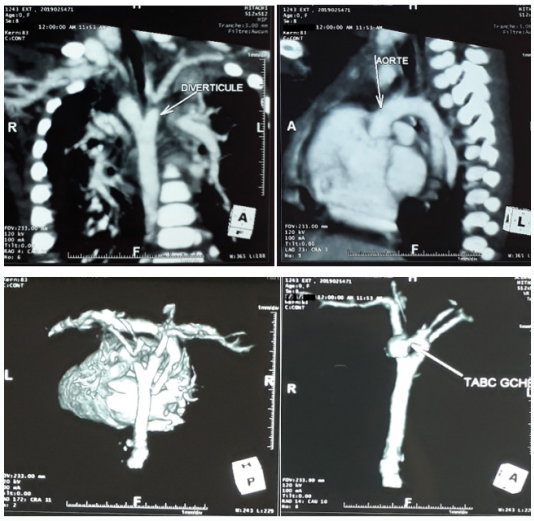

Figure 3: Thoracic CT angio showing a double aortic arch.

Cardiac ultrasound revealed a heart of normal architecture, structure and function with a right aortic arch.

Surgical cure of the non-functioning aortic arch was indicated and the infant was operated on at the age of 6 months; he died one month later due to postoperative complications.

Case 3

A female infant, 2.5 months of age, hospitalized for respiratory difficulties, from a non-consanguineous marriage and a well-monitored pregnancy carried to term. The delivery was vaginal, Apgar 9/10 and the birth weight 3000g. Her vaccination is up to date according to the Expanded Programme on Immunisation (EPI). His psychomotor development was normal. On admission he had a weight of 5.4 kg, a temperature of 37°C, a heart rate of 140 beats/min, a heart rate of 50 cycles/min and an SpO2 of 95% (room air). The pleuropulmonary examination noted respiratory distress with signs of struggle and bilateral ronchi. The rest of the examination was normal.

The blood count was normal: white blood cells = 8400/mm^3, haemoglobin = 11.9g/dl, platelets = 246,000/mm^3; CRP was negative (<5 mg/l).

Chest X-ray showed hyperclarity of the lungs, lowering of the diaphragmatic cupolas and horizontalization of the ribs.

Cardiac ultrasound was normal. The hypothesis of acute bronchiolitis was evoked. The infant received oxygenation, nebulised β2 mimetics and corticosteroid therapy.

As the clinical signs persisted, a CT scan was ordered. It showed a double aortic arch with a dominant right arch giving rise to the brachiocephalic trunk, while the smaller left arch gives rise to a left brachiocephalic trunk. The 2 trunks join to form the descending aorta. The aerodigestive axis (trachea and oesophagus) is enclosed between the two arches, with extrinsic stenosis of the trachea, the diameter of which measures 2 mm at this level (Figure 4).

The infant was operated on at the age of 9 months with identification of the dominant arch and suture section of the other arch allowing release of the oesotracheal tract. The postoperative course was simple and uncomplicated.

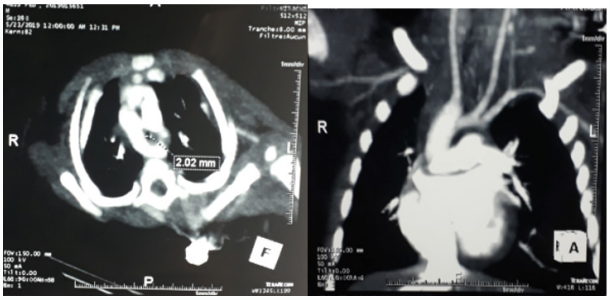

Figure 4: Chest CT scan showing a double aortic arch with right arch dominance.

Discussion

The aortic arch is a complex structure, derived from the development of six successive arches between the aortic sac and the dorsal aortas. Remodelling of these arches produces in 70% of cases a left aortic arch giving 3 branches: the brachiocephalic trunk, the left common carotid artery and the left subclavian artery. Benign variations are common and of no consequence. However, more complex variations, such as the double aortic arch, can produce tracheo-oesophageal compression. The age of onset varies from the neonatal period to childhood depending on the severity of the oesophageal compression and the existence of associated malformations. In our series, we had an early diagnosis in all patients before the age of 7 months, which is consistent with the data found in the literature. The double aortic arch is due to the non-regression of the fourth right aortic arch, which causes the ascending aorta to divide in front of the trachea into two arches that pass around the trachea and oesophagus to enclose them and then join behind the oesophagus to form the descending thoracic aorta [5]. In our series, the right arch was dominant in all 3 observations, the left arch was non-permeable in observation 2. The circumstances of discovery of a double aortic arch are variable, it may be discovered during the work-up of a cardiac disease or during a radiography made in a systematic way [6], but most often in the newborn and the infant it is the signs of oesophageal compression that attract the attention as it was the case in our 3 observations [2,7]. Clinically the syndrome is characteristic with stridor, episodes of respiratory distress and sometimes asphyxia [7,8]. Signs of dysphagia are often unrecognised and in the background. Stridor may be enhanced by tracheomalacia and may also result in an asthmatic syndrome [3,5,9]. In our series, respiratory signs were found in all patients, but digestive signs were present in only one case. Stridor was observed in the first case. Respiratory infections are the most common finding [10]. They were found in our series. The double aortic arch may be isolated or associated with congenital heart disease. The most frequently associated heart diseases are: tetralogy of Fallot, ventricular septal defect, patent ductus arteriosus, open septal pulmonary atresia, coarctation of the aorta [11,12]. Echocardiography cannot always allow the diagnosis of a double aortic arch to be made with certainty, other examinations such as angiography-CT or MRI must be used, but it plays an essential role in the search for any associated congenital heart disease. In our series, all patients underwent transthoracic echocardiography and returned without abnormality. Aortic angio-CT using the spiral technique is the examination of choice for a complete and exhaustive morphological evaluation of the anomalies of the aortic arches and of any associated congenital heart disease. It confirms the diagnosis of DAA and specifies the relationship of the aortic arch with the oesophagus, trachea and the position of the supra-aortic trunks, serving as a preoperative mapping [12,13]. Treatment of the double aortic arch is surgical. Surgical repair of the double aortic arch was defined in 1945 by Gross [14]. The postoperative course is generally simple with complete disappearance of symptoms [2,15] as was the case in two of our patients. However, complications such as chylothorax, laryngeal oedema, sepsis, haemorrhagic complications, persistence of respiratory manifestations, notably stridor, may be reported [2].

Conclusion

Double aortic arch is a rare congenital malformation. It is the most common cause of symptomatic complete vascular ring. Therefore, a tracheal or oesotracheal compression syndrome in the neonatal period and in infants should always lead to a search for an anomaly of the aortic arches and in particular the double aortic arch.

References

1. Patrick Olivier MYERS De, Meyrin. The Aortic Arch Embryology, Anatomy & Anatomical Variants for the Clinician. [Geneva]: Faculty of Medicine, University of Geneva; 2009.
2. Ouedraogo AR, Boncoungou K, Maïga S, Ouedraogo G, Badoum G, Diallo O, et al. A case of aortic arch malformation simulating asthma. Rev Pneumol Clin. 2018;74:253-6.
3. Lemouakni S, Hassani A, Elyajouri H, Kmari M, Ourrai H, Abilkacem R, et al. Double aortic arch: about two cases. Pan Afr Med J. 2017;27:273.
4. Ammar S, Belhassen S, Laamiri R, Sahnoun L, Ksia A, Ezzi A, et al. P-025 - Diagnostic and surgical management of double aortic arches. Arch Pediatrics. 2015;22:239.
5. Breviere Gm. Anomalies of the aortic arches. Encycl. medchir (éditions scientifiques et médicales Elsevier sas. Paris all rights reserved) cardiology, 11- 940- d- 80, 2003; 9p.
6. Kouki S, Fadhel A, Landoulsi M, Boujemaa H, Ben Abdallah N. CT atlases of anatomical variations of the supra-aortic trunks. Feuill Radiol. 2015;55:133-45.
7. Lincoln JCR, Deverall PB, Stark J, Aberdeen E, Waterston DJ. Vascular anomalies compressing the oesophagus and trachea. Thorax. 1969;24(3):295-306.
8. Backer CL, Ilbawi MN, Idriss FS, DeLeon SY. Vascular anomalies causing tracheoesophageal compression. Review of experience in children. J Thorac Cardiovasc Surg. 1989;97(5):725-31.
9. Ouedraogo AR, Boncoungou K, Maïga S, Ouedraogo G, Badoum G, Diallo O, et al. A case of aortic arch malformation simulating asthma. Rev Pneumol Clin. 2018;74(4):253-6.
10. Alsenaidi K, Gurofsky R, Karamlou T, Williams WG, McCrindle BW. Management and Outcomes of Double Aortic Arch in 81 Patients. Pediatrics. 2006;118(5):e1336-41.
11. Kastler B, Livolsi A, Germain P, Bernard Y, Michalakis D, Rodiere E, et al. Contribution of MRI in the exploration of congenital heart and large vessel anomalies. J Radiol. 2004;85:1821-50.
12. Oguz B, Haliloglu M, Karcaaltincaba M. Paediatric multidetector CT angiography: spectrum of congenital thoracic vascular anomalies. Br J Radiol. 2007;80(953):376-83.
13. Amina K. THE DOUBLE AORTICAL ARC (About 11 cases). [Morocco]: Faculty of Medicine of FES; 2015.
14. Gross RE. Surgical relief for tracheal obstruction from a vascular ring. N Engl J Med. 1945;233:586-90.
15. Sarbeji M, Azevedo I, de Blic J, Le Bourgeois M, Scheinmann P, Paupe J. Vascular arch abnormalities in asthmatic children. Rev Fr Allergol Immunol Clin. 1991;31:35-40.