**Scholars Journal of Medical Case Reports**
Abbreviated Key Title: Sch J Med Case Rep
ISSN 2347-9507 (Print) | ISSN 2347-6559 (Online)
Journal homepage: https://saspublishers.com

OPEN ACCESS

**Radiology**

# Double Aortic Arch: A Case Report

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**DOI:** 10.36347/sjmcr.2021.v09i11.011 | **Received:** 31.09.2021 | **Accepted:** 07.11.2021 | **Published:** 12.11.2021

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**Abstract** **Case Report**

The double aortic arch represents a rare anomaly of the aortic arch. It results from the lack of involution of the dorsal caudal aorta. Clinical symptomatology is usually early, noted in the neonatal period or shortly thereafter, dominated by respiratory and digestive signs. TOGD allows an accurate diagnosis of the anomaly. However, angiography is of great diagnostic interest as well as in the choice of the therapeutic approach. Only surgical treatment can remove the compressive signs on the trachea-esophageal axis. The objective of our work is to illustrate through an observation of double aortic in an infant aged 7 months, the contribution of imaging in the difficult diagnosis of this anomaly.

**Keywords:** Double aortic arch - respiratory distress - angioscan - bronchiolitis - congenital - aortic button.



## INTRODUCTION

Double aortic arch is a rare anomaly of the aortic arch. It results from the lack of involution of the dorsal caudal aorta [1]. Clinical symptomatology is usually early, noted as early as the neonatal period. Angioscan is of great diagnostic interest as well as in the choice of therapeutic approach. Only surgical treatment allows the removal of compressive signs on the trachea-esophageal axis. The operative mortality has become low thanks to the progress of postoperative resuscitation. We report through this observation of double aortic arch, the contribution of imaging in the difficult diagnosis of this anomaly.

## PATIENT AND OBSERVATION

### Observation

Male infant, 7 months old, from a poorly monitored pregnancy, delivered vaginally in hospital, immediate cry. Had a history of hospitalization at 2 days of age for first episode of acute viral bronchiolitis. Admitted to the pediatric department for etiological management of his repeated respiratory infections. Clinical examination found a polypneic infant, apyretic, sibilant rales on auscultation, with signs of respiratory struggle. A chest X-ray showed thoracic distension, absence of aortic button with a cardiothoracic index of 0.5 (Figure 1). The paraclinical workup was completed by a thoracic angioscan showing two aortic arches, right and left, surrounding the trachea and esophagus before joining to the descending thoracic aorta, each providing the origin of the primary carotid artery and the homolateral subclavian artery, both crosses are permeable with a predominance of the right arch, there are associated to a condensation of the middle lobe and Fowlers sitting of an air bronchogram with bronchial micronodules of the two pulmonary fields of infectious appearance (Figure 2,3) with a normal cardiac ultrasound.

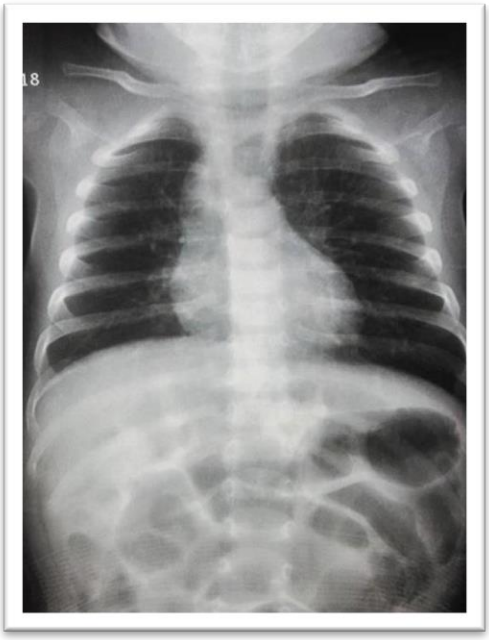

**Fig-1: Chest X-ray showing thoracic distension with effacement of the aortic button**

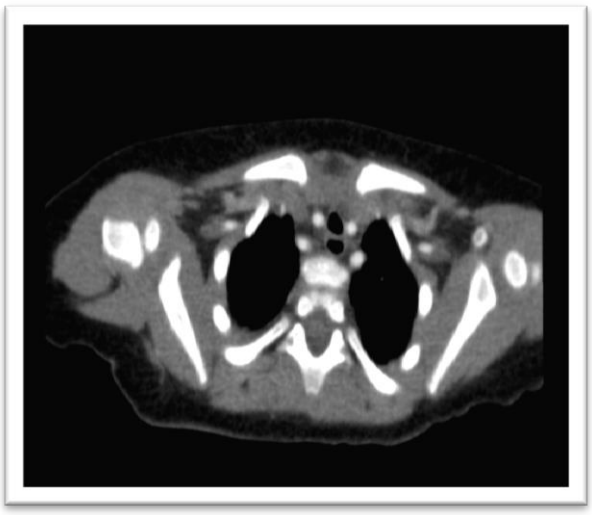

**Fig-2: Axial scan section showing the origin of the carotid and subclavian arteries of both aortic arches**

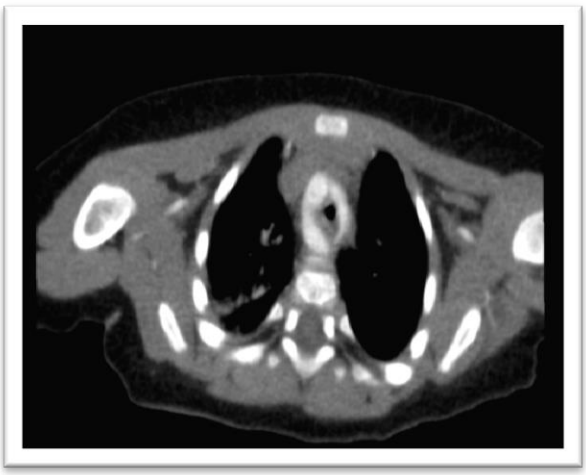

**Fig-3: Axial CT section showing a double aortic arch encircling the oeso-tracheal axis**

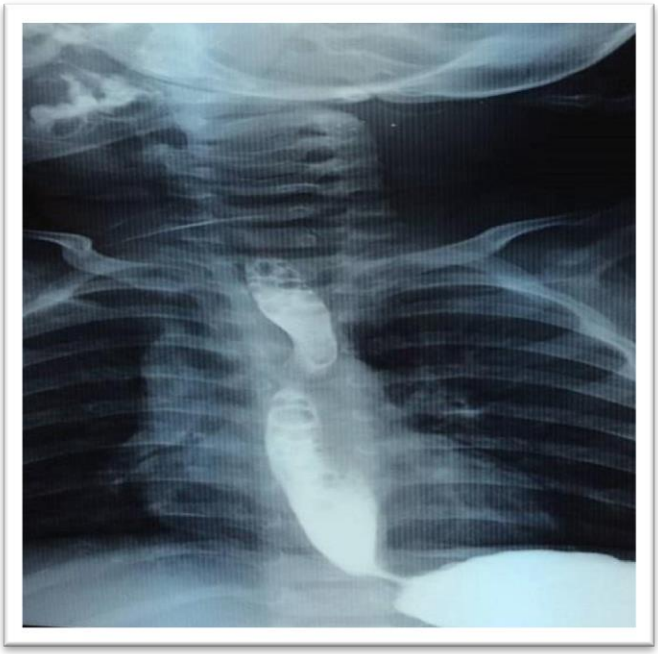

**Fig-4: Esophageal transit showing an impression of the esophagus**

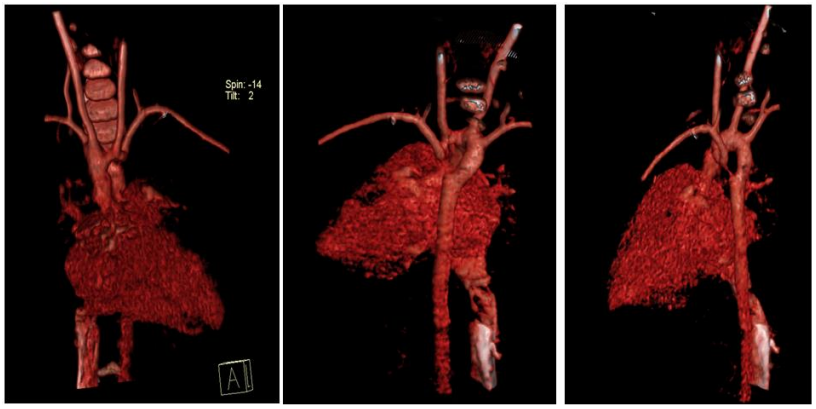


**Fig-5: 3D demonstration of the double aortic arch (posterior, anterior and profile)**

## DISCUSSION

Anomalies of the aortic arches have been known since the 18th century. Their anatomical description was first described in 1737 by Hommel, then their radiological aspects in 1926 by Arkin and those of the retroesophageal right subclavian in 1936 by Kommerell. It was not until 1939 that the clinical description of double arches by Wolman was realized [13]. Aortic arch anomalies have a common embryological origin: they may involve the aorta, the vessels of the aortic root or the pulmonary arteries. They are rare and account for 1% of cardiovascular congenital anomalies [13, 14]. The double aortic arch anomaly is the most common and accounts for 40-50% of symptomatic vascular rings. It is due to the persistence of the distal portion of the right dorsal aorta, resulting in the formation of right and left aortic crosses that surround the trachea and esophagus before joining to form the descending thoracic aorta. Each arch provides the origin of the primary carotid artery and the homolateral subclavian artery. Usually, both junctions are patent, in which case the right or left may be dominant or they may be equal in size. In 75% of cases the right arch is dominant according to the literature [2-5] as in the case of our patients. The age of onset of the disease was early in our two patients (7 months, 9 months), which seems to be consistent with the literature which is usually before 3 years of age [1], although in 25% of cases the diagnosis is made in adulthood [6]. Double aortic arch can be difficult to diagnose because the symptoms are not typical of a cardiac disorder. The clinical picture is dominated by respiratory (stridor, respiratory distress, chronic cough) and digestive (dysphagia) symptoms [7, 8]. The importance of the signs depends on the space between the two aortic arches. Sometimes the diagnosis can be confused with asthma, bronchiolitis or recurrent pneumonia. This congenital vascular anomaly can be isolated or associated with cardiac malformations such as tetralogy of Fallot, ventricular septal defect, patent ductus arteriosus, open septal pulmonary atresia, coarctation of the aorta, or extracardiac malformations such as microdeletion of chromosome 22q11 in Di George syndrome. Cardiac ultrasound of our patients came back normal. Imaging plays a major role in the diagnosis of double aortic arch. Chest radiography is the first line of investigation. Fibroscopy is used to determine the degree of compression and to eliminate differential diagnoses. The oeso-gastro-duodenal transit sometimes allows, depending on the location and orientation of the impression, to specify the type of anomaly. Any symptomatic child with an abnormal esophageal impression on TOGD should have cross-sectional imaging (Figure 4). Angioscan is the reference examination; it allows making the diagnosis, to specify its type and its relationship with the adjacent structures. Currently, thanks to new technology, an angioscan with reconstruction gives an excellent image in two or three dimensions (Figure 5) [9, 10]. MRI, like CT, allows a precise anatomical assessment to be made; it is not irradiating and is reputed to be non-invasive, although it requires heavy sedation. Treatment is exclusively surgical [10-12]. The indication for surgery is indicated if the oeso-tracheal compression syndrome is severe with recurrent respiratory infections and episodes of asphyxia which may be complicated by cardiorespiratory arrest. The left posterolateral thoracotomy is the reference route for the different authors. An experienced surgical team is required with good anesthetic preparation.

## CONCLUSION

The double aortic arch remains a rather rare pathology that it is necessary to think of it in front of a neonatal respiratory distress without obvious etiology, in front of a severe Asthma of the infant or bronchiolitis with repetition. Surgical treatment is indicated if the symptomatology is frank and/or life-threatening. Generally, the postoperative evolution is satisfactory. A karyotype is requested to search for micro deletion 22q11.

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