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The Double Aortic Arch: About Two Cases and Literature Review

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Abstract: The double aortic arch is a rare embryological anomaly; it constitutes < 1% of all congenital heart defects. It leads to the formation of a complete vascular ring encircling the tracheo-esophageal axis, causing respiratory and digestive symptoms. Clinical presentation is often variable, delaying diagnosis. The treatment is mainly surgical, aimed at lifting compression on the tracheo-oesophageal tract. We report two observations, in order to illustrate the authenticity and the particularity of this anomaly in clinical, therapeutic and prognosis terms.
Keywords: double aortic arch, embryological anomaly.

INTRODUCTION

The double aortic arch is a complete vascular ring [1]. The term vascular ring is used to describe malformations of the aortic arch that cause cerclage of the esophagus and trachea [2], which is responsible for a variable symptomatology. The usual cause is an inadequacy, during embryonic life, of the normal regression of the aortic arches, which are born from the arterial trunk [2]. The double aortic arch is an anomaly most often isolated and rare (7% to 17%) [3, 4]. The diagnostic approach is conditioned by the circumstances of discovery. Surgical treatment consists in lifting the compression of the tracheo-oesophageal axis, this is done in the majority of cases by left thoracotomy, with extensive release of the various elements of the mediastinum [1].

The prognosis of this anomaly is related to functional abnormalities in tracheomalacia type, and anatomical abnormalities in trachea type hypoplasia, which then significantly increase the burden of management [1].

We report this observation, in order to illustrate the authenticity and particularity of this anomaly both on the clinical, therapeutic and prognosis level.

CASES REPORTS

Case-1:

This refers to the 9-month-old infant with a history of recurrent wheezing dyspnea requiring multiple visits and the notion of repeated bronchial infection.

During the clinical examination, the patient was conscious with good staturo-ponderal and psychomotor development. The auscultation had objectivized sibilants in both pulmonary fields.

The patient was hospitalized in the paediatric ward, where he was given a chest x-ray with objective evidence of diffuse bronchial syndrome (Figure-1).

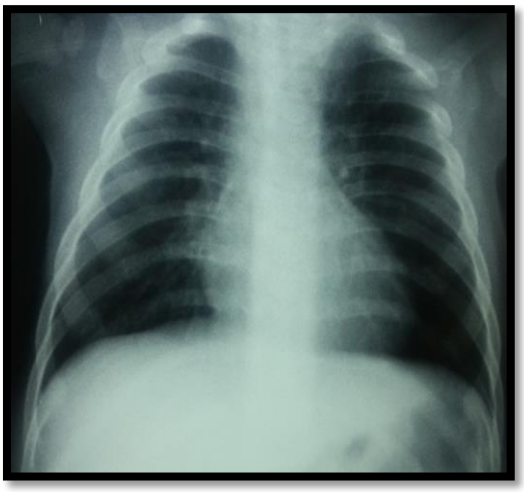

Fig-1: chest x-ray

And a chest CT scan without and with contrast medium injection allowed diagnosis of an anomaly of the aortic arches with a double aortic arch type with a left atresic arch responsible for stenosing tracheal compression (Figure-2).

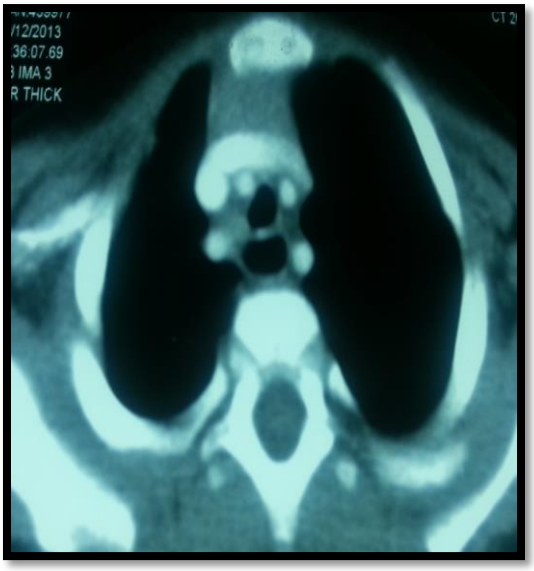

Fig-2: a chest CT scan with contrast medium injection

The patient was transferred to the cardiovascular surgery department for surgical management, a trans-thoracic echorgaphy was performed during the pre-therapeutic check-up, without any associated abnormalities.

The surgical procedure consisted of resection of the left arch, under general anesthesia, after monitoring of the invasive blood pressure at the right femoral artery, NIBP, SaO2, ECG, bladder catheterization.

Left posterolateral thoracotomy in the 3rd intercostal space. Careful dissection and placement in view of the descending aorta followed by persistent arterial canal, distal parts of the aortic arches and the left subclavian artery, while preserving the left recurrent laryngeal nerve (Figure-3 & 4).

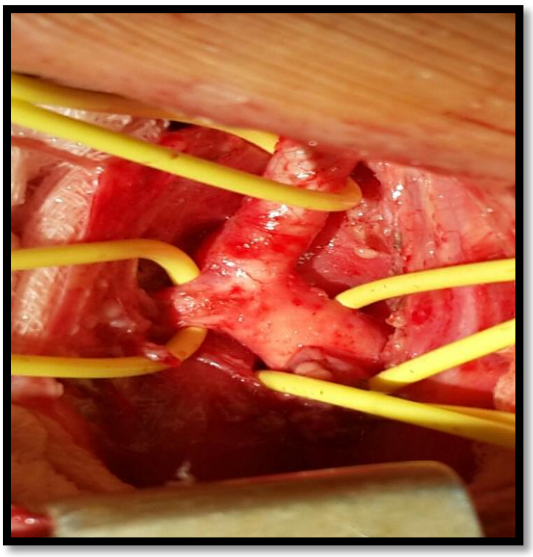

Fig-4: perioperative view of the left subclavian artery, ascending aorta and the left arteric arch.

Accidental section of the thoracic canal that was closed by two clips. The distal posterior part is larger in diameter than the anterior. The permeability of the latter was evaluated by intermittent clamping on both sides of the birth of the left subclavian artery under monitoring of the saturation curve on the left upper limb. It is decided to ligate the distal left arch upstream of the birth of the left subclavian artery (Figure-5 & 6).

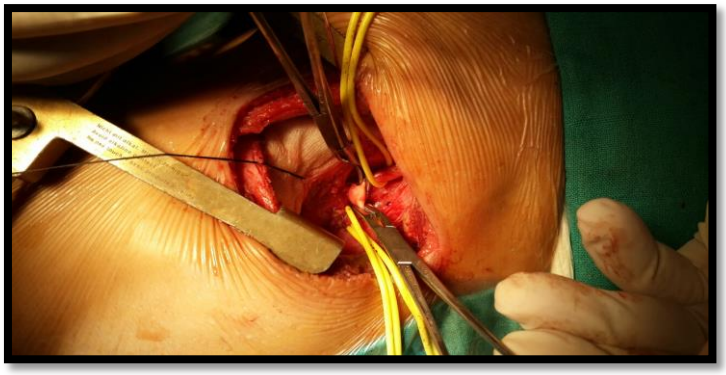

Fig-5: Ligation of the left distal arch upstream of the birth of the left subclavian artery

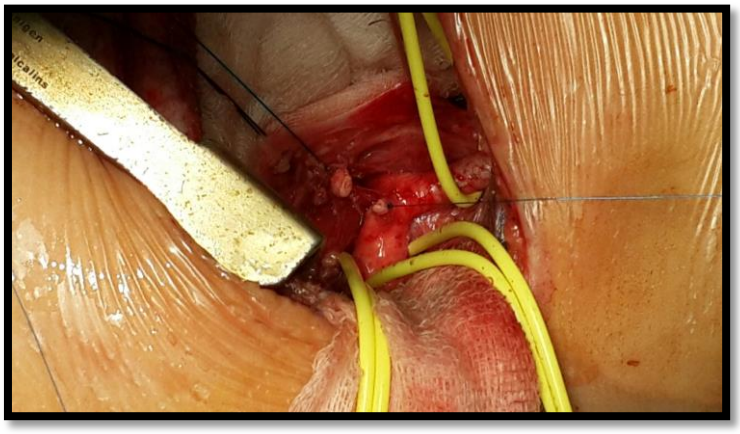

Fig-6: Double ligation and persistent arterial canal section

Aortoplexy from the descending aorta to the prevertebral fascia is performed. The procedure is completed by extensive dissection of the adhesions surrounding the part opposite the trachea and on either side of the esophagus.

The patient was transferred to the cardiovascular resuscitation unit. Post-operative follow-up was simple.

Case-2:

This refers to infants aged 1 year and 09 months, resulting from a marriage without consanguineity, with no specific pathological history. During the interogation, the mother reported stress dyspnea, suffocating cough and recurrent dysphagia since the age of 1 year.

During the clinical examination, the patient was conscious with good staturo-ponderal and psychomotor development. The auscultation had objectivized snoring Rales at both lung fields.

Chest x-ray and trans-thoracic echocardiography are normal (Figure-7). The positive diagnosis of a double aortic arch was made by the thoracic scanner with injection of contrast material (Figure-8).

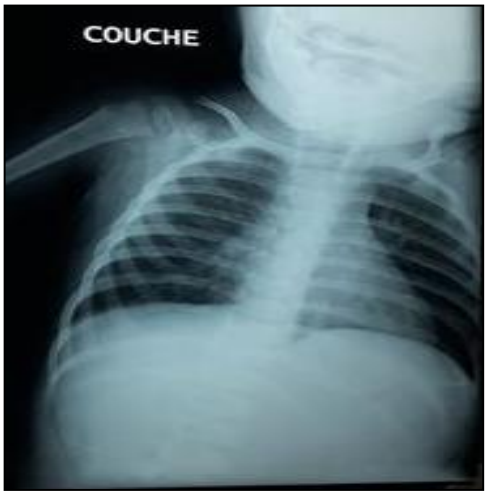

Fig-7: Chest x-ray of the face

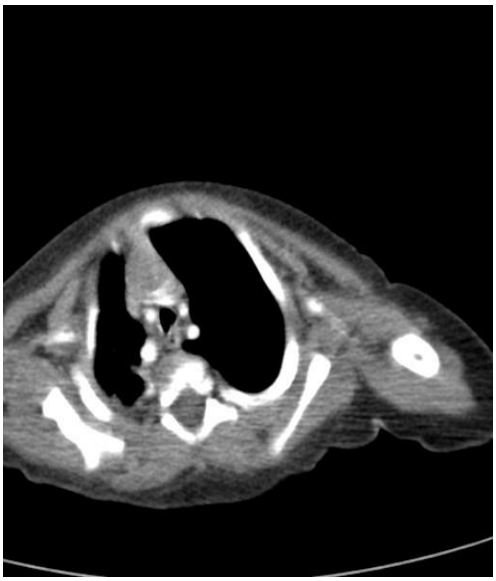

Fig-8: Injected thoracic scanner for diagnosis of the double aortic arch

The surgical procedure consisted of resection of the left arch, under general anesthesia:

Posterolateral left posterolateral thoracotomy at the 3rd intercostal space. Thorough dissection and placement on lakes of the descending aorta and then the persistent arterial canal, distal parts of the aortic arches and the artery under the left clavicle, while preserving the left recurrent laryngeal nerve (Figure-9).

The posterior arch (right) is dominant prolonged by the descending aorta, the anterior arch (left) dominated is atresic at its distal part (closing the ring) under tension by an arterial ligament, and printing the anterior left face of the aerodigestive axis.

The permeability of each arch was evaluated by intermittent clamping on both sides of the birth of the left subclavian under monitoring of the saturation.

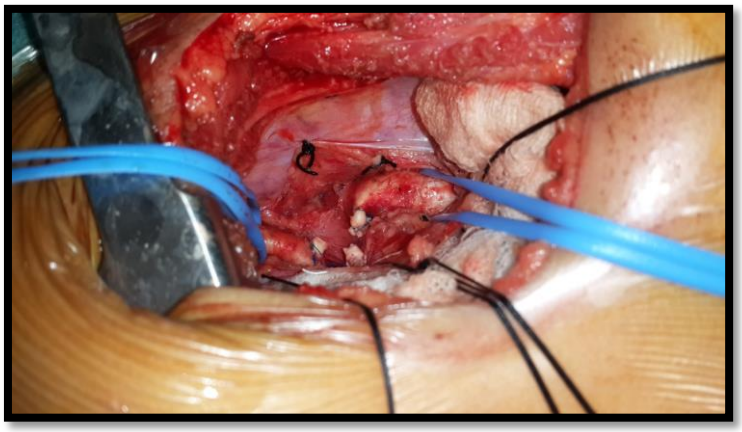

Fig-9: Ligature and section of the left arch near its origin in the subclavian artery

It was decided to cut the distal left arch downstream of the birth of the left subclavian artery. Ligation/section and suture of the persistent arterial canal

The procedure is completed by extensive dissection of the adhesions surrounding the part opposite the aerodigestive axis.

DISCUSSION

The double aortic arch is the most frequent malformation of the vascular ring which results in a strapping of the tracheo-esophageal axis. The double aortic arch is defined anatomically as a complete vascular ring, which is formed by two equal right and left aortic arches, or a right or left atretic arch [2].

The double aortic arch is normally isolated, but it can be associated with other congenital heart diseases with conotroncular malformations, respiratory and digestive manifestations can be hidden by the most striking signs of cyanosis and left heart failure which are very common during these heart diseases.

Our patient had a double aortic arch with a dominate right arch, which is the most frequent variant (> 70%) [1], the carotid artery and the right subclavian artery are born from the right aortic arch, while the carotid artery and the left subclavian artery are born from the left aortic arch separately.

Usually, the diagnostic orientation is guided by interrogation and chest X-ray, which allows to visualize a narrowing of the localized and fixed tracheal caliber, an anomaly of position of the aortic button [1]. Fibroscopy can be useful in the diagnosis of the double aortic arch, in order to eliminate a laryngeal or subglottic anomaly, the degree, location and vascular nature of compression, and can also objectify a tracheomalacia, which conditions the prognosis [1].

The oeso-gastroduodenal transit was mentioned as the most valuable means of investigation, and of first intention for the diagnosis of the double aortic arch [5, 6] by highlighting an esophageal notch, but this examination remains insufficient for the preoperative evaluation of the patient [7].

The advent of new imaging techniques by CT and cardiac MRI, approved their superiority in diagnosis and preoperative assessment, specifying the vascular anatomy, its relationship with trachea and esophagus [2] and constituting a very reliable alternative to the oeso-gastroduodenal transit.

Trans-thoracic ultrasound can confirm the diagnosis of double aortic arc, and is also very useful in detecting other associated congenital heart disease. The subcostal section of the left ventricle ejection is used to visualize the aortic bifurcation by starting from the aortic valve, this generates a Y-bifurcation, centripetal, and an encircling morphology of the tracheo-oesophageal axis. A major disadvantage of subcostal cutting is that the area of interest is confined to far-field imaging, which can be given a reduced resolution in larger patients [8].

The sternal fork approach can overcome this problem in imaging the area of interest in the near field. Through a large axis / sagittal plane of the sternal fork (with left obliquity) and a 30 degree counterclockwise rotation of the transducer (with right obliquity), we can visualize a left and right arc, respectively [8].

Then, a small axis/coronal plane of the sternal fork can be used to visualize the double aortic arc using a scanning plane [8]. The conclusion of an AAD diagnosis by performing only a small axis/coronal view sternal fork is risky, due to the large number of false positive images that are caused by aliasing the right phantom pulmonary artery as a vascular ring [8]. The major problem with this examination is its limited

ability to clearly imagine the structures of the anterior aortic arch and arterial ligament.

Although cardiac catheterization and angiography may provide a diagnosis of AAD and differential diagnosis of rare types of aortic arc abnormalities, and pulmonary slings, and canal slings, but catheterization is neither systematic not necessary, especially in small patients with severe cardiorespiratory disorders [9, 10].

The surgical indication is based on a tripod that combines clinical symptomatology, a vascular malformation recognized at the CT scan, tracheal compression of more than 50% with fibroscopy [1].

Thoracotomy should be performed on the side of the arterial aortic arch and on the side of the arterial ligament persistence, compression of the trachea and esophagus is relieved by ligation and division of the small arch and persistent arterial canal. We prefer performed a left thoracotomy and divide the left arch just below the origin of the subclavian artery then divide the ligament, thus releasing the esophagus and trachea. After the operation, the patient recovered relief from his stridor and dysphagia [2].

In patients with isolated double aortic arc, surgical splitting of the lower aortic junction is indicated. However, pre-operative recognition of associated abnormalities is important, especially in cases of coarctation or atresia of one or two arches, the main congenital heart diseases associated with tracheomalacia and bronchomalacia. on the one hand, the surgical division of an aortic arch without pre-operative coarctation recognition or an atresia of the other arch, which makes post-operative follow-up difficult and sometimes fatal [8].

In addition, tracheomalacia can be complicated by tracheomalacia, with tracheal edema sometimes requiring prolonged intubation, and narrowing of the trachea and esophagus may persist for a long time [1].

Finally, preoperative bronchoscopy can be used to exclude tracheomalacia or associated bronchomalacia, which conditions the patient's short- and long-term prognosis [7].

Overall mortality is 3.3 to 4.3% depending on the series with a darker prognosis if congenital heart disease associated, while survival at 35 years and 96%; 75% without respiratory functional sign at 1 year of surgery [1].

CONCLUSION

The double aortic arch is a rare pathology, responsible for early clinical signs, most often. The Surgical treatment involves lifting the compression of the tracheo-oesophageal axis. The prognosis is essentially related to its functional complications, and the complexity of anatomical anomalies.

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