Cardiology in the Young (2014), **24**, 534–536
doi:10.1017/S104795111300070X

Brief Report

Double aortic arch: postnatal obliteration of the left aortic arch. Is arterial duct closure responsible?

David Rose,[1] Riccardo D'Ascoli,[1] Flavia Ventriglia,[2] Fabio Miraldi[1]

[1]*Department of Cardiocirculatory Pathophysiology, Anesthesiology, and General Surgery;* [2]*Pediatric Cardiology, Sapienza University of Rome, Rome, Italy*

Abstract We present a case of double aortic arch with a predominant right and a double arterial duct detected by echocardiogram in a 28-week gestation foetus. The first evaluation revealed that both arches were perfused; the 1-month postnatal echocardiogram showed the closure of both arterial ducts and the partial obliteration of the left aortic arch between the left subclavian artery and the dorsal aorta. In our case, the postnatal obliteration of the left arch in a double aortic arch was probably due to the closure of the left-sided arterial duct.

Keywords: Double aortic arch; arterial duct; vascular ring

Received: 22 February 2013; Accepted: 11 April 2013; First published online: 29 May 2013

A DOUBLE AORTIC ARCH, AS DESCRIBED IN 1964 BY Edwards,[1] represents a persistence of both right and left fourth aortic arches that form a complete vascular ring around the trachea and oesophagus, which rarely, by encirclement and compression of mediastinal structures, may produce symptoms of airway obstruction and dysphagia, requiring surgery.[2] Arterial duct or ligament is often involved in vascular rings, but probably is also involved in their pathogenesis. We recently encountered a patient with a double aortic arch in whom the closure of the left duct might have led to obliteration of the left aortic arch.

Case report

In a 28-week gestation foetus, a prenatal echocardiogram revealed a double aortic arch with a predominant right and a double arterial duct. Both arches were perfused (Fig 1a). The 1-month postnatal echocardiogram showed the closure of both arterial ducts and the partial obliteration of the left aortic arch between the left subclavian artery and the dorsal aorta (Fig 1b). The infant became progressively symptomatic for dysphagia and stridor on inspiration. Physical examination was normal. The patient underwent a barium esophagogram, which demonstrated a deep persistent extrinsic indentation in the posterior aspect of the oesophagus, consistent with the diagnosis of double aortic arch. Computed tomography scan confirmed the diagnosis, and it was no longer possible to detect flow through the left aortic arch (Fig 2a). The patient was operated at 4 months of age, and the surgical procedure, through a left 4th intercostal space thoracotomy, consisted of the resection of both the left aortic arch remnant, ligament, close to the dorsal aorta, and the left arterial ligament (Fig 2b). The post-operative course was uneventful and the patient was discharged on post-operative day 4.

Discussion

Double aortic arch and the right arch with an aberrant subclavian artery are the two most common forms of vascular ring, a class of congenital anomalies of the aortic arch system in which the trachea and oesophagus are completely encircled by connected segments of the aortic arch and its

Correspondence to: D. Rose, Department of Cardiocirculatory Pathophysiology, Anesthesiology, and General Surgery, Sapienza University of Rome, 00161 Rome, Italy. Tel: +00 393 207 260 389; Fax: +00 390 649 977 2599; E-mail: davidrose@libero.it

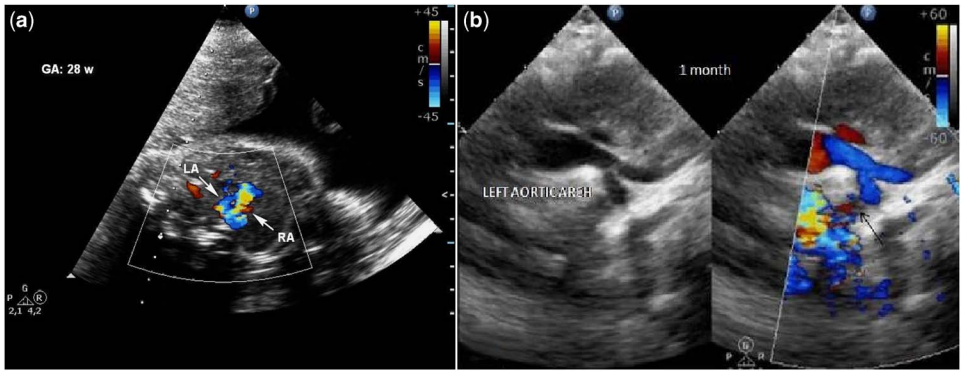

Figure 1.
*(**a**) Prenatal echocardiography at gestational age of 28 weeks revealing a double aortic arch with a predominant right and a double arterial duct, both perfused. (**b**) Postnatal echocardiography performed at 1 month showing the closure of both arterial ducts and the partial obliteration of the left aortic arch between the left subclavian artery and the dorsal aorta (black arrow). LA = left arch; RA = right arch.*

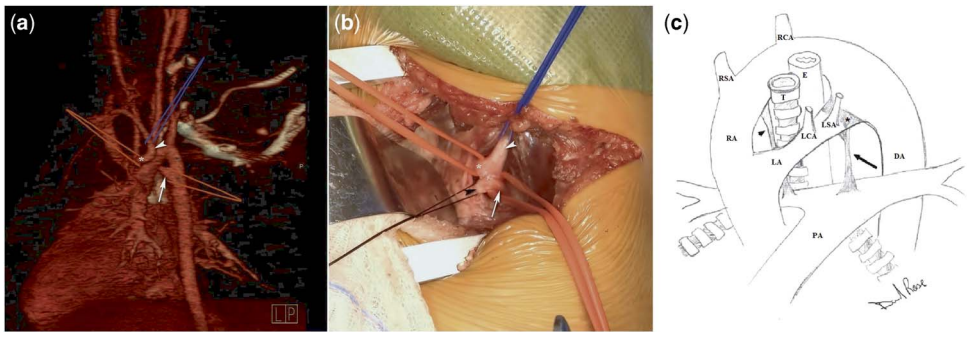

Figure 2.
*(**a**) CT scan. Asterisk: left aortic arch between left carotid artery and left subclavian artery. White arrow: remnant of left aortic arch post left subclavian artery. Tip of white arrow: left subclavian artery. (**b**) Intraoperative aspect. Asterisk: left aortic arch between left carotid artery and left subclavian artery. White arrow: remnant of left aortic arch post left subclavian artery. Tip of white arrow: left subclavian artery. Tip of black arrow: left arterial ligament. (**c**) Schematic representation of the vascular ring. Tip of black arrow, right arterial ligament. Black arrow, left arterial ligament. Asterisk, obliterated left arch after left subclavian artery. DA = descending aorta; E = esophagus; LA = left arch; LCA = left carotid artery; LSA = left subclavian artery; PA = pulmonary artery; RA = right arch; RSA = right subclavian artery; RCA = right carotid artery; T = trachea.*

branches.[1] Usually, this anomaly is associated with right arterial ductus patency.[3]

We describe a postnatal obliteration of the left arch in a double aortic arch probably due to the closure of the left-sided arterial duct. This combination of events created a symptomatic vascular ring: predominant right aortic arch, a left obliterated aortic arch, ligament, and a double-sided arterial ligament.[4] In this patient, we had patency of both left and right arterial ducts during the prenatal period; the postnatal closure of both of them had different effects on the anatomy of the two arches.[5] The underlying mechanisms responsible for these changes probably are the same described for the aetiology of the aortic coarctation. The two theories are: the ductal tissue theory and reduced-flow theory. In the former, tissue from the arterial duct invades the aorta, and when it obliterates it results in an aortic constriction. In the latter, the defect can develop secondary to haemodynamic changes that reduce flow to the involved vessel.[6] Embryogenesis of vascular rings is interpreted as the consequence of the reabsorption or obliteration of different segments of the paired original pattern of aortic arches.[7] The ring caused by double ducts/ligaments, left aortic arch fibrous continuity in a right aortic arch in the present case is considered of interest because the obliteration of the mentioned segment occurred in infancy, starting from a classic double aortic arch pattern.

Acknowledgment

The authors would like to thank Professor Pietro Gallo for his precious advise.

https://doi.org/10.1017/S104795111300070X Published online by Cambridge University Press

Financial Support

This research received no specific grant from any funding agency, commercial or not-for-profit sectors.

Supplementary materials

For supplementary material referred to in this article, please visit http://dx.doi.org/10.1017/S104795111300070X

References

1. Edwards JE. "Vascular rings" related to anomalies of the aortic arches. Mod Concepts Cardiovasc Dis 1948; 17: 19–20.
2. Yoo SJ, Min JY, Lee YH, Roman K, Jaeggi E, Smallhorn J. Sonographic diagnosis of aortic arch anomalies. Ultrasound Obstet Gynecol 2003; 22: 535–546.
3. Kanne JP, Godwin JD. Right aortic arch and its variants. J Cardiovasc Comput Tomogr 2010; 4: 293–300.
4. Craatz S, Künzel E, Spanel-Borowski K. Right-sided aortic arch and tetralogy of Fallot in humans – a morphological study of 10 cases. Cardiovasc Pathol 2003; 12: 226–232.
5. Thankavel PP, Brown PS, Lemler MS. Left-dominant double aortic arch in critical pulmonary stenosis and ventricular septal defect. Pediatr Cardiol 2012; 33: 1469–1471.
6. Liberman L, Gersony WM, Flynn PA, Lamberti JJ, Cooper RS, Stare TJ. Effectiveness of prostaglandin E1 in relieving obstruction in coarctation of the aorta without opening the ductus arteriosus. Pediatr Cardiol 2004; 25: 49–52.
7. Lee M-L, Chen M, Tsao L-Y, et al. Congenital stridor and wheezing as harbingers of the del22q11.2 syndrome presenting cardiovascular malformations of right aortic arch, aberrant left subclavian artery, Kommerell's diverticulum, and left ligamentum arteriosum. Cardiovasc Pathol 2011; 20: 124–129.