Double aortic arch with hypoplastic right aortic arch and type C atresia of left aortic arch

Luca Costanzo, Elio Caruso, Salvatore Agati and Paolo Guccione*

Department of Pediatric Cardiology and Cardiac Surgery, Bambino Gesù Children’s Hospital Centro Cardiologico Pediatrico del Mediterraneo, San Vincenzo Hospital, Taormina, Italy

* Corresponding author. Department of Pediatric Cardiology and Cardiac Surgery, Bambino Gesù Children’s Hospital Centro Cardiologico Pediatrico del Mediterraneo, San Vincenzo Hospital, Contrada Sinna 98038 Taormina (Messina), Italy. Tel: +39-06-68593913; fax: +39-06-7593540; e-mail: paolo.guccione@opbg.net (P. Guccione).

Received 11 February 2014; received in revised form 2 April 2014; accepted 9 April 2014

Abstract

We report the case of a 2-month-old baby with a double aortic arch, type C atresia of the left arch and severe hypoplasia of the right aortic arch between the right carotid and subclavian arteries, resulting in systemic obstruction, left ventricular dysfunction and congestive heart failure. Surgical augmentation of the right aortic arch ameliorated the obstruction with improvement in left ventricular function and symptoms.

Keywords: Double aortic arch • Aortic arch variations • Cardiac failure

INTRODUCTION

Double aortic arch (DAA) is an unusual congenital malformation resulting from abnormal regression of parts of the aortic arches originating from the truncus arteriosus during foetal life [1].

DAA is classified into 2 types depending on the patency of the 2 arches: Type 1, where both arches are functioning, and Type 2, where 1 of the arches is atretic; Type 2 is further classified into 4 subtypes (A, B, C and D) depending upon atresia location [2].

We present a case of DAA with hypoplastic right aortic arch (RAA) and type C atresia of the left aortic arch (LAA).

CASE REPORT

A 2-month-old female baby with a negative family history of congenital heart disease was referred to our hospital because of heart murmur, poor feeding, increasing dyspnoea, tachycardia and pallor. A 3/6 systolic murmur was heard at the apex. Multiple haemangiomas over the right side of the face, mouth and right retroauricular regions were detected. Transthoracic echocardiogram revealed: left ventricular (LV) dysfunction (ejection fraction 30%), LV dilatation (LV diastolic diameter 35 mm, Z-score + 5.78), LV hypertrophy (8 mm, Z-score + 3.52), severe mitral regurgitation and normal origin of coronary arteries. Conversely, a suboptimal suprasternal window showed an abnormal RAA with a peak systolic pressure gradient of 88 mmHg and diastolic run off at Doppler evaluation.

Cardiac catheterization revealed a peak gradient of 70 mmHg between the ascending and thoracic aorta, and LV end-diastolic pressure was 19 mmHg. Angiography showed cervical RAA with severe hypoplasia (minimum diameter 1.5 mm) of the segment between the right common carotid artery (RCCA) and the right subclavian artery (RSA) (Fig. 1A and B). RCCA was hypoplastic with multiple obstructions and irregular course. The LAA was interrupted after emergence of the left common carotid artery (LCCA), which was the only unobstructed vessel emerging from the ascending aorta. The flow in the right vertebral artery was inverted and multiple collateral branches towards the RSA were noted. Retrograde injection into the descending aorta revealed a remnant of LAA from which a dysplastic left subclavian artery (LSA) and a small patent ductus arteriosus (DA) originated. Balloon angioplasty of the RAA was attempted and partially relieved the obstruction (Fig. 1C and D).

Computed axial tomography and 3D reconstruction confirmed the complex form of DAA (Fig. 1E-G). Furthermore, multiple areas of vascular enhancement were found in the right submandibular and retroauricular regions. A drawing depicting the peculiar anatomy of the DAA is shown in Fig. 1H.

The baby underwent surgical repair of the hypoplastic RAA. The operation was performed by means of median sternotomy. Normothermic partial left CPB was established through cannulation of the ascending and descending aorta. The RAA was clamped proximally after the emergence of the LAA and distally above the diaphragm level of the descending aorta. The RAA was incised longitudinally from the cervical hypoplastic segment to the descending aorta and reconstruction was completed with a pulmonary artery homograft augmentation patch. After careful dissection, DA was easily identified and excised but the remnant of the LAA could not be identified with certainty because of the unusual anatomy and LAA section was not performed. After surgery, the invasive gradient decreased from 55 to 15 mmHg.

The postoperative period was uneventful and the infant was easily weaned from mechanical ventilation. Ventricular function
Figure 1: Angiography showed cervical right aortic arch (RAA) with RAA hypoplasia in frontal (A) and lateral projection (B); result after balloon dilatation in frontal (C) and lateral projection (D). Three-dimensional reconstruction of CT scan in front (E), posterior (F) and lateral view (G). Drawing depicting the peculiar anatomy of the double aortic arch found in our patient (H). *Narrowest segment of hypoplastic right aortic arch; DAo: descending aorta; LAA: left aortic arch; LCCA: left common carotid artery; LSA: left subclavian artery; LVA: left vertebral artery; RCCA: right common carotid artery; RSA: right subclavian artery; RVA: right vertebral artery.

Figure 2: Multiple haemangiomas. (A and B) The haemangiomas before cardiac surgery; (C and D) progressive reduction in size 18 months after surgery.
and symptoms rapidly improved. Recurrence of obstruction of the reconstructed RAA was detected by echocardiography at 2 and 8 months after surgery and successfully treated by balloon angioplasty. After 18 months, the patient was asymptomatic, and her growth percentile improved from the 20th to 35th percentile. LV function normalized. Facial haemangiomas progressively decreased and disappeared (Fig. 2).

DISCUSSION

DAA is an anomaly caused by failure of regression, or regression at an abnormal site, of the fourth aortic arch [3]. DAA can be associated with segmental atresia of one of the arches classified into 4 subtypes according to the location of the atretic segment: types A and B are relatively common, but types C and D are rare [4].

We reported a case of DAA with hypoplastic RAA and LAA type C atresia. Our patient presented with congestive heart failure because of severe systemic obstruction, whereas the most commonly reported clinical presentation of DAA is caused by airway compression [1, 5].

Our surgical strategy was to reconstruct the aortic segment with an augmentation patch because the hypoplastic segment of the RAA was long and tortuous and direct anastomosis between the ascending and descending aorta was advised against; in addition, the marked cervical position of the RAA clearly contraindicated thoracotomy. Notably, surgical dissection to identify the remnant of LAA was interrupted to avoid any damage to the left laryngeal recurrent nerve, the phrenic nerve and the LSA. The decision not to excise LAA was also supported by the fact that the patient was not symptomatic for stridor or feeding disorder and the cervical position of the RAA made the area between the RAA and LAA sufficiently wide.

In our case, the potential development of collateral circulation was limited by hypoplastic RCCA and LSA and the origin of RSA being distal to the obstruction. Hence, the LCCA was the only patent supraaortic vessel that could supply the collateral circulation towards the RSA. Thus, distal aortic perfusion was guaranteed retrogradely from the intracranial vertebrospinal arterial system and through multiple extracranial superficial collateral branches. Therefore, we postulated that the multiple haemangiomas localized exclusively on the right side of the head could be the expression of such a collateral pathway. Indeed, these vascular anomalies progressively disappeared after improvement in the RAA obstruction.

With such complex DAA presentations, echocardiography might reveal the underlying cause of cardiac dysfunction but cardiac catheterization, angiography and CT scan provide a precise anatomical and physiological evaluation, allowing the establishment of an appropriate therapeutic strategy.

Conflict of interest: none declared.

REFERENCES


eComment. Right cervical aortic arch and aberrant left subclavian artery

Author: Jose Aramendi
Hospital de Cruces, Bilbao, Spain
doi: 10.1093/icvts/ivu198
© The Author 2014. Published by Oxford University Press on behalf of the European Association for Cardio-Thoracic Surgery. All rights reserved.

We read with interest the report on double aortic arch with hypoplastic right aortic arch and type C atresia of the left aortic arch [1]. The anatomy described is that of a right cervical aortic arch with retro-oesophageal left subclavian artery. However, this pathology is not a true double aortic arch although it looks like it, as the segment between the left carotid and left subclavian arteries is absent. Only in those rare cases where a fibrotic ligament connecting those arteries exists could we consider a double aortic arch. In fact, the authors could not identify the left aortic arch during surgery, simply because it did not exist, and there was no vascular ring, which is the main characteristic feature of double aortic arch. The concept of double aortic arch with partial atresia of the left aortic arch is no longer in use and the classification in type 1 and 2 plus subtypes A to D is too complex [2]. Nearly all surgeons are now using simple descriptive terms to describe these vascular rings rather than these antiquated classification schemes. In particular, I would refer the authors to the nomenclature and database article [3]. We published a similar case of right aortic arch with retro-oesophageal left subclavian artery, left ductus arteriosus and anomalous origin of right pulmonary artery from the ascending aorta [4]. Our case did not have arch stenosis but required ductus ligation and section plus right pulmonary artery reimplantation.

Conflict of interest: none declared.

References