CASE REPORT – CONGENITAL

One-stage repair of aberrant left brachiocephalic artery and coarctation of the aorta in right aortic arch

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Abstract

The combination of right aortic arch with aberrant left brachiocephalic artery and aortic coarctation is very rare. Here, we report the case of a 3.1-kg neonate with multiple malformations who received detailed preoperative anatomical definition by chest computed tomography (CT) scan and eventually underwent one-stage repair at the age of 17 days. The surgical technique included left brachiocephalic artery detachment and its end-to-end anastomosis to the branch of a monofurcated pulmonary homograft used to treat the aortic arch coarctation simultaneously. Postoperative control chest CT scan performed 3 months postoperatively revealed optimal repair. Considerations on imaging and surgical technique are reported.

Keywords: Vascular ring · Aberrant left brachiocephalic artery · Right aortic arch

INTRODUCTION

The combination of right aortic arch with aberrant left brachiocephalic artery and aortic coarctation is very rare. To the best of our knowledge, only 2 such cases have previously been reported in the literature [1, 2]. Thus, according to the classification suggested by Nis and Vidne [3] for coarctation with the right aortic arch, this should be the third case of Type D abnormality reported in the literature, and probably the first describing in detail the surgical technique adopted.

CASE REPORT

A 3.1-kg neonate with multiple malformations (palatoschisis, choanal atresia, facial dysmorphism, right eye coloboma and cryptorchidism) suggestive of Charge syndrome came to our attention for respiratory symptoms associated with hypersalivation. The patient underwent routine investigations, including chest X-ray, two-dimensional (2D) echocardiography, body and head ultrasound scan and oesophageal transit study, suggesting a vascular ring with almost complete obstruction of the oesophagus. Two-dimensional echocardiography showed a complex and unclear right aortic arch anomaly with a 45-mmHg mean Doppler gradient suggestive of coarctation. On cardiovascular computed tomography (CT) scan of the chest, performed by iodine contrast agent (Iopamiro 370) intravenous injection with a Dual Source Multislice CT scanner (Siemens Somatom Definition Flash, Forchheim, Germany), aortic coarctation and vascular ring were detected, the latter consisting of a right aortic arch with the oesophageal left innominate artery arising from the descending aorta through a Kommerell’s diverticulum and an anterior left-sided arterial ligamentum that completed the ring (Fig. 1A). Focusing on the aortic arch anatomy, the site of coarctation was located at the level of the transverse arch between the right carotid artery and right subclavian artery, with a minimum diameter of 2 mm. A non-restrictive atrial septal defect coexisted. Surgery was performed on the 17th day of life, and one-stage repair was undertaken through midline sternotomy coexisitent with hypothermic cardiopulmonary bypass. During cooling, the aberrant left innominate artery was clamped at its origin from the descending aorta, transected, and taken anteriorly to the oesophagus and trachea after thorough dissection. Then, under circulatory arrest, the transverse aortic arch was widely incised longitudinally to reach the site of the left innominate artery detachment distally and the origin of the right carotid artery proximally (Fig. 2A). A monofurcated cryopreserved homograft was trimmed to simultaneously create a patch and leave the pulmonary branch untouched (Fig. 2B). The patch portion of the homograft was then sutured to the aortic incision using 7-0 polypropylene, and aortic arch reconstruction was achieved. The branch portion of the homograft was used to fill the gap between the arch and the transected left innominate artery and their end-to-end anastomosis was performed with a running 7-0 polypropylene suture (Fig. 2C). During rewarming, the atrial septal defect was closed through right atriotomy, and the aortic clamp was released after a 50-min cardiopulmonary arrest time. Cardiopulmonary bypass was discontinued with the patient in sinus rhythm, and the sternum was closed on the second postoperative day. The early postoperative course was smooth, but the patient could not be extubated for the tracheomalacia demonstrated at fibroscopy. Performing a support tracheostomy allowed for disconnection from the ventilator on the 34th day.
postoperative day and discharge home in good clinical condition on the 104th postoperative day. Since discharge, the child has been feeding and thriving well, with no clinical signs of oesophageal compression. Three months after discharge he underwent both 2D echo and chest CT scan investigations, showing good aortic arch morphology (Fig. 1B), with no aortic gradient and normal flow in the left carotid artery.

**DISCUSSION**

Generally, coarctation in the right aortic arch encompasses both complex obstructive lesions and anomalous branching of brachiocephalic vessels. The embryological development of this anatomical setting is well elucidated by the Edward’s hypothetical double aortic arch model \[4\], with persistence of the right fourth branchial arch and regression of the ventral segment of the left fourth branchial arch proximal to the left carotid artery.

The surgical technique described here was based on the use of a patch of homologous tissue with a branch for direct reimplantation of the left aberrant innominate artery. As an alternative, we could try to use native tissue exclusively by repairing the arch by sliding plasty and directly reimplant the aberrant left innominate artery after mobilization. We felt more confident to rely on the described technique in order to avoid possible stretching produced by direct reimplantation of an anomalous vessel arising too distal and posterior to the oesophagus.

In the experience reported, 2D echocardiography was not completely efficient in exploring the distal aortic arch and the initial segment of the descending aorta. A CT scan performed by a new generation multislice scanner with fast acquisition provided detailed vascular and airway anatomy, allowing high-quality three-dimensional (3D) volume rendering reconstruction. We wish to emphasize the crucial role of this diagnostic tool for planning the optimal surgical treatment of vascular rings, particularly for the more complex anatomical shapes.

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**Figure 1:** (A) Preoperative CT scan 3D reconstruction showing the right-sided aorta with a high aortic arch at the level of chest aditus. The left brachiocephalic trunk is located posteriorly, behind the oesophagus, and its origin from the descending aorta comes from a Kommerell diverticulum. (B) Postoperative CT scan 3D reconstruction showing unobstructed aortic arch repair and unstretched reimplantation of the left innominate artery.

**Figure 2:** Reconstructive technique. (A) Preoperative view. RSA: right subclavian artery; RCA: right common carotid artery; RVA: right vertebral artery; LBA: left brachiocephalic artery; LCA: left common carotid artery; LVA: left vertebral artery; *dotted line = site of detachment of the LBA, **dotted line = longitudinal incision of the aortic arch. (B) Trimming of the monofurcated pulmonary homograft to simultaneously create a patch and leave the left pulmonary branch untouched. (C) Final result. *dotted line = site of reimplantation of the LBA, **dotted line = aortic arch reconstruction with monofurcated homograft patch (see text).
Conflict of interest: none declared.

REFERENCES


eComment. Rare combination of aortic anomalies

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We have read with great interest the article by Gandolfo et al., in which they shared their strategy for one-stage repair in a patient with Charge syndrome, an aberrant left brachiocephalic artery and coarctation of the aorta in the right aortic arch. It is a well-structured case report study. Recently, we had a similar patient with a right-sided hypoplastic aortic arch, aberrant left subclavian artery, Kommerell’s diverticulum and a retroaortic course of the innominate vein.

Our patient was a 1.5-year-old girl referred to our clinic for surgical correction of hypoplasia of the aortic arch. She was diagnosed with PHACE (posterior fossa malformations, haemangiomas, arterial anomalies, cardiac defects, eye abnormalities) syndrome after birth. She had no history of dysphagia or dyspnoea. After the incidental finding of a heart murmur, transthoracic echocardiography (TTE) was performed. The pressure gradient measured in the descending aorta was only 60 mmHg. No additional cardiac abnormalities were found. Cardiac catheterization showed a right-sided aortic arch with an aberrant left subclavian artery. The entire aortic arch was hypoplastic. Moreover, it was shown that the left subclavian artery had aberrant course originating from a region distal to the left common carotid artery with diverticulum of Kommerell in its isthmus. To better understand the morphology, a computed tomography scan was performed and in addition to those anomalies, retroaortic course of the left innominate vein was identified. Preoperative findings were consistent with intraoperative observations.

PHACE syndrome is an uncommon association between large infantile haemangiomas, usually of the face, and structural abnormalities of the brain, cerebral vasculature, eyes, and/or aorta [1]. The association of coarctation of the aorta/hypoplasia of aortic arch and right-sided aortic arch is uncommon in non-syndromic patients [2]. An anomalous course of the innominate vein is rare. Right-sided aortic arch may have been with retroaortic course of innominate vein and more than 80% of the patients with anomalous left innominate vein have obstruction of the right ventricular outflow tract (RVOT), commonly tetralogy of Fallot (TOF) with or without pulmonary atresia [3, 4]. However, as in our case, combination of right-sided aortic arch with hypoplasia, aberrant left subclavian artery and retroaortic course of innominate vein without RVOT obstruction has not been published yet. In contrast to simple aortic coarctation or aortic arch interruption observed in non-syndromic patients, aortic anomalies found in the context of PHACE and other syndromes, such as Charge syndrome may represent a distinctive morphological entity with unusually complex and unpredictable anatomic involvement. For that reason, accurate interpretation of the anomaly as well as the related anatomical structures is crucial, especially in patients with syndromes.

Conflict of interest: none declared

References


