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*

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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.

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**References**