

A successful surgical correction for a rare aortic arch interruption

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Introduction: Aortic arch interruption (AAI) is a rare congenital heart defect. Blood flow to the descending aorta is exclusively dependent on the ductus arteriosus making survival without treatment rare after the neonatal period. In rare cases AAI is associated with right-sided descending aorta and in these cases it is usually associated with DiGeorge syndrome. Treatment is surgical with anastomosis of both aortic ends.

Clinical case: We present the case of a newborn whose parents were first cousins. Full-term pregnancy and childbirth without complications. The newborn was admitted to a Neonatal Intensive Care Unit on day one of life for cyanosis. A diagnose of *ostium secundum* atrial septal defect (ASD), perimembranous ventricular septal defect (VSD) and type A aortic arch interruption (distal to the origin of the left subclavian artery) with left-sided aortic arch and right-sided descending aorta was made. DiGeorge syndrome was suspected and later confirmed. An angio-TC was performed that confirmed the echocardiographic findings and showed that the distance between the left-sided distal end of the aortic arch and the right-sided proximal end of the descending aorta was 15mm, making an anatomic correction technically challenging. Corrective surgery was performed at 11 days of life. The descending aorta was anastomosed to the posterior wall of the ascending aorta, proximal to the emergence of the supra-aortic trunks, and the ASD and VSD were closed. Time of extra-corporeal circulation was 2 hours and 34 minutes. There were no significant post-operative complications. Three months after surgery the infant is doing well. The aorta has laminar blood flow with 10 mmHg of maximal pressure gradient in the neo-aortic isthmus.

Conclusion: In AAI with left-sided aortic arch and right-sided descending aorta anatomic correction is technically challenging. The anastomosis of the descending aorta to the ascending aorta overcame this problem with good surgical and clinical outcome.

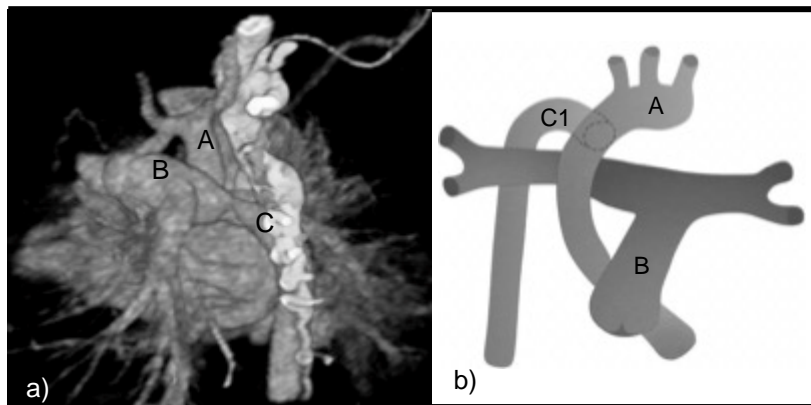


Fig. 1: Preoperative and post-operative anatomy - a) Preoperative thoracic angio-TC image, b) Scheme of the postoperative anatomy.
A: aortic arch, B: main pulmonary artery, C: descending aorta, C1: descending aorta anastomosed to the posterior face of the ascending aorta.

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