Cardiology in the Young

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Double trouble: double aortic arch with bilateral interruption and bilateral arterial ducts

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Case Report

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Abstract

Double aortic arch with bilateral interruptions is a rare abnormality. Indeed, to the best of our knowledge, it has not previously been reported before. Rare variants, such as a double arch with ligamentous atresia of the left component and double arch with bilateral coarctation, have received previous attention. These rare examples can pose significant diagnostic challenges for those relying on transthoracic echocardiography. Use of cardiac CT or MRI is now critical for prompt surgical planning and correction.

Introduction

Malformations of the aortic arch, or arches, comprise approximately 3% of congenital heart defects.[1] These malformations can appear independently or together with other intracardiac defects and chromosomal abnormalities.[1–3] Clinically, such anomalies may be asymptomatic. They are then often discovered incidentally during imaging for other indications.[1–3] Alternatively, they may manifest with symptoms caused by tracheal compression, such as neonatal respiratory distress, wheeze, stridor, recurrent infections, and dysphagia, or oesophageal compression resulting in feeding difficulty, dysphagia, or vomiting when the anatomy produces vascular rings.[1–3]

We present a rare case of a double aortic arch with bilateral interruptions and bilateral persistence of the arterial duct in a patient with a 22Q11 deletion. The double aortic arch is itself the consequence of bilateral persistence of the arteries of the fourth pharyngeal arches, which encircle the trachea and oesophagus to create a vascular ring.[2–4] The interruption is the consequence of bilateral regression of parts of both arterial arches.[2,5] Examples of double arches are well described with ligamentous atresia of various segments, which cannot be readily visualised on postnatal echocardiography, cardiac CT, or MRI.[4,6] These anomalies can pose significant challenges for a timely diagnosis and intervention.[6] A case series has been collected of double arches with ligamentous atresia of the left arch distal to the left subclavian artery, with the point made that the combination can easily be confused with a right aortic arch with mirror-imaged branching.[6] This emphasised that patients with suspected malformations of the aortic arches require high-resolution imaging for correct diagnosis and subsequent accurate surgical decision-making.[1–3]

Case

We present a case of a female infant who is currently 10 months old and who was born at term at an outside hospital with no complications. The mother had received routine obstetric care including a prenatal ultrasound scan, expressed no significant concerns during pregnancy, and did not undergo fetal echocardiogram. The child was cyanotic at birth. An echocardiogram revealed a large posterior malalignment ventricular septal defect, a moderately hypoplastic aortic valve (Figure 1), and an interrupted aortic arch. Prostaglandin was initiated immediately, and she was transferred to our hospital for surgical management. Subsequent echocardiograms were suggestive of isolated subclavian arteries supplied by bilateral arterial ducts, a large ventricular septal defect, and suspicion of bilateral interruption of a double aortic arch (Video clip 1). Cardiac CT (Figure 2, Video clip 2) showed that the proximal aortic arch branched to become bilateral common carotid arteries but was then interrupted on both sides. The descending aorta was located on the right, with bilateral large and tortuous arterial ducts feeding both subclavian arteries. Genetic testing was positive for 22Q11 deletion, and the infant also had a partial cleft palate.

The patient underwent placement of bilateral bands on the right and left pulmonary arteries at 1 month of age. At 6 weeks of age, she underwent a Norwood operation, with repair of the

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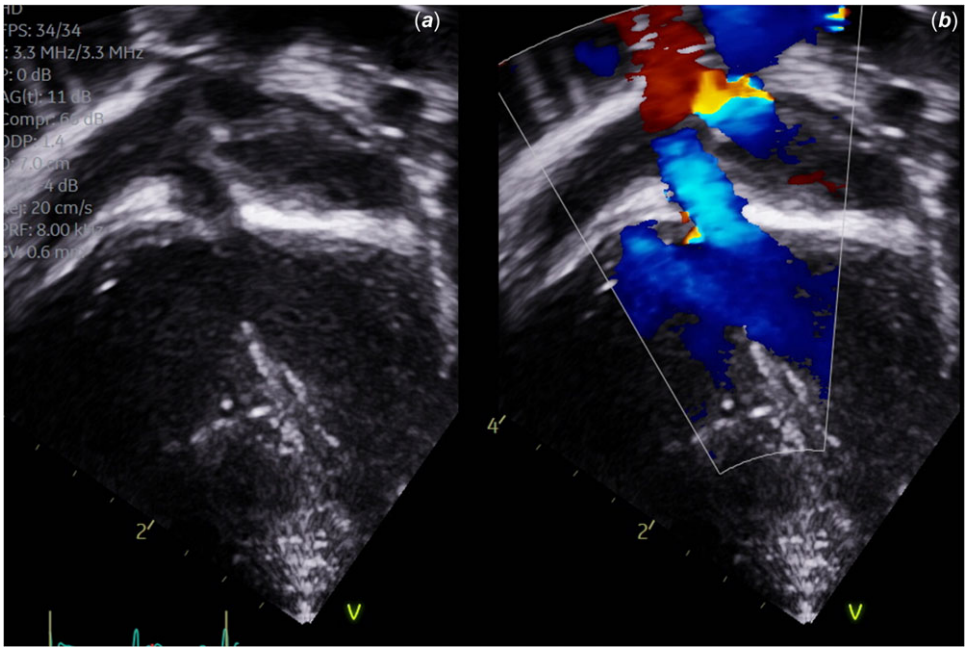


Figure 1. Panel a shows a two-dimensional echo image of a hypoplastic aortic valve and a large posterior malalignment ventricular septal defect. Panel b shows the colour Doppler of the aortic valve and ventricular septal defect.

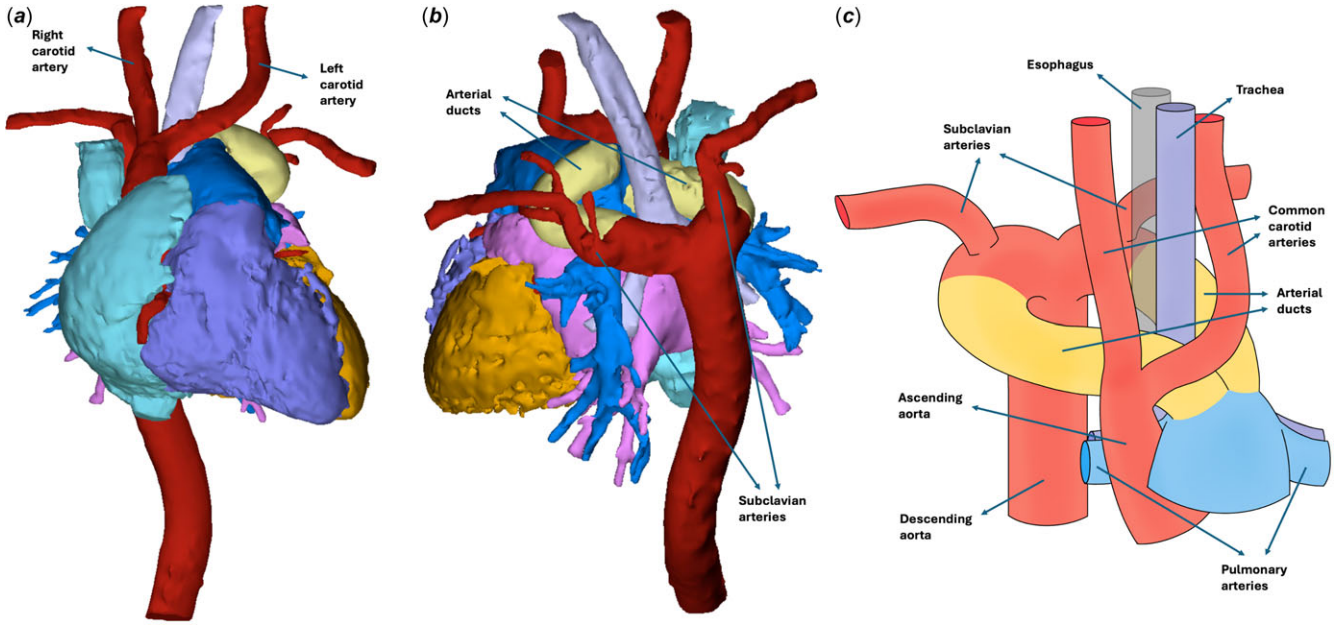


Figure 2. Three-dimensional CT reconstruction of the double aortic arch with bilateral interruption and bilateral arterial ducts shown from anterior aspect (panel a), posterior aspect (panel b), and simplified hand drawing (panel c).

interrupted aortic arch, Damus–Kaye–Stansel anastomosis, placement of a 5-millimetre shunt from the right ventricle to pulmonary artery, removal of pulmonary arterial bands, and division of the left arch remnant behind the oesophagus, with translocation of the left subclavian artery to the persisting patent right-sided aortic arch. During the surgical procedure, it was confirmed that the hypoplastic ascending aorta gave rise to the left and right common carotid arteries. The left-sided arterial duct arose superiorly from the pulmonary trunk, gave rise to the left subclavian artery, and then coursed medially behind the oesophagus and trachea to join the descending aorta rightward of the midline. The right-sided arterial duct arose rightward from the pulmonary trunk just superior to the right pulmonary artery and coursed laterally, giving off the right subclavian artery before passing to join the right-sided descending aorta. The left-sided duct and the retro-oesophageal part of the left arch were mobilised, transfixed, ligated, and divided just distal to the origin of the left subclavian artery, thereby removing any subsequent potential for oesophageal compression. The right subclavian artery was left attached to the proximal descending aorta, and after resecting all the right distal ductal tissue, the interrupted arch was repaired by anastomosing the descending aorta to the posterior aspect of the ascending aorta, with a small patch of pulmonary homograft used to augment the anastomosis. The transected pulmonary trunk was

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then anastomosed directly to the ascending aorta without the use of a patch.

She subsequently developed stenosis of the right ventricle to pulmonary artery shunt and the left pulmonary artery, requiring balloon angioplasty and stent placement in the left pulmonary artery at 4 months of age. The most recent operation, undertaken at 8 months of age, was upsizing the shunt to 7 millimetres and left pulmonary artery patch augmentation. Her most recent CT with three-dimensional reconstruction showed widely patent right-sided aortic arch, unobstructed shunt, and mildly hypoplastic central pulmonary artery with good-sized distal branch pulmonary arteries (Video clip 3). She is currently doing well with oxygen saturations of 80–85% with 0.5 litres per minute of nasal cannula oxygen. She has a normal heart rate and is gaining weight in an adequate fashion. Her last echocardiogram showed no significant residual lesions and normal biventricular systolic function.

Discussion

To our knowledge, this is the first reported case of interruption of both components of a double aortic arch with bilateral persistence of arterial ducts.[1] Prior reports have described cases of double aortic arch with bilateral aortic coarctation diagnosed by invasive angiography, surgical examination, echocardiography, and CT.[1] In our case, serial echocardiograms and CT findings confirmed the diagnosis in a clinically symptomatic patient. Aortic arch anomalies are also frequently detected incidentally during imaging for CHD and other conditions, underscoring the need for a systematic visual assessment of all cardiac structures in these patients.[1,2]

Multiple imaging modalities can be used to delineate the anticipated complex anatomy and branching of the components of double arches, along with their relationships to the trachea and oesophagus and associated abnormalities.[2,3] "Conventional modalities such as barium esophagography, chest radiography, and catheter angiography are falling out of practice due to difficulties in their use in children and their limitation in providing only two-dimensional images.[2,3] Echocardiography is the prevailing first-line test in neonates and is facilitated by the large thymic acoustic window. It is operator-dependent, however, and creates difficulties in visualising posterior vascular structures. It is unable to quantify tracheal or oesophageal compression produced by vascular rings."[1–3,6] Because of these caveats, CT or MRI are preferred techniques for providing detailed evaluation of the vascular morphology and providing three-dimensional reconstructions to confirm the diagnosis and plan surgery.[1–3] Use of MRI avoids radiation but may require longer acquisitions and anaesthesia in infants and can be limited by claustrophobia.[2,3] CT angiography often offers the best balance of high spatial resolution, minimal need for anaesthesia, and breath-holding. Modern low-dose radiation protocols make it particularly attractive in paediatrics.[2,3,6]

In summary, a patient with suspected malformations of the aortic arch should receive serial, segmental echocardiography followed by CT imaging to confirm the diagnosis and inform surgical planning.[1–3,6] Prompt and appropriate imaging is critical to avoid late diagnosis and delayed surgery, which can lead to chronic issues caused by tracheal and oesophageal compression.[1–3,6]

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