Single institution experience with a right-sided interrupted aortic arch

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Abstract

Interrupted aortic arch is a rare congenital heart disease, whereas right-sided interrupted aortic arch is an extremely rare disorder with few reported cases in the literature. We report our institutional experience with two such patients and review the recent literature.

Keywords: Right-sided interrupted aortic arch • Microdeletion 22q11 • Modified Yasui procedure • Magnetic resonance angiography • Computed tomography angiography

CASE SERIES

Patient 1

A full-term female weighing 3.27 kg was referred due to poor perfusion and desaturations 24 h after birth. An echocardiogram showed a right-sided interrupted aortic arch (IAA) type B, large posterior malaligned ventricular septal defect (VSD), small secundum atrial septal defect, bicuspid aortic valve, aortic valve annulus measured at 7 mm and right-sided ductus arteriosus. These findings were confirmed by cardiac catheterization. The patient initially remained on prostaglandins and underwent an 8-mm interposed graft placement between the ascending and descending aorta, which the surgeon chose based on his assessment at the time of the surgery, main pulmonary artery banding via right thoracotomy and ductal ligation on the 7th day. Staged palliation was chosen for this patient due to the unusual arrangement of the great vessels, even though our institutional practice had changed to primary complete repair of IAA in 2000. Her genetic work-up was positive for the DiGeorge syndrome. She underwent staged repair at 4 months of age that included Dacron® patch closure of VSD, polytetrafluoroethylene (PTFE) patch closure of the atrial septal defect, PTFE patch aortoplasty and main pulmonary artery debanding. She developed significant right ventricular outflow tract obstruction related to the previous band and pulmonary valve adhesion to the VSD patch and underwent transannular patch repair 3 years after the initial surgery (Fig. 1). She continued to do well and underwent an additional 16-mm interposed graft that was chosen by the surgeon during the operation and was placed via the right chest through the right side of the pericardium, as the ascending aorta could not be approached via a median sternotomy due to the presence of the oesophagus directly anterior to the descending aorta, because of a left pulmonary artery repair at the age of 4 years. On her recent follow-up, the patient remained asymptomatic with mild supravalvar aortic and pulmonary stenosis related to her previous surgeries.

Patient 2

A 2-week-old full-term infant girl weighing 3.2 kg presented to us with cyanosis. The child was started on prostaglandins at presentation and required mechanical ventilation with inotropic support. An echocardiogram was performed upon arrival that showed a right-sided IAA type B with bilateral carotid arteries arising from the ascending aorta and bilateral subclavian arteries coming off the descending aorta distal to the ductal insertion, bicuspid aortic valve (valve annulus = 3.5 mm, z-score ~ -6.6), large posterior malaligned VSD and hypoplastic ascending aorta that measured 6 mm (z-score ~ -3.5). The patient was hypocalcaemic at presentation and later tested positive for 22q11 chromosomal microdeletion. A magnetic resonance angiography was performed that confirmed the echocardiographic findings (Fig. 2). Following stabilization, she underwent a modified Yasui procedure via a median sternotomy that included a Damus-Kaye-Stansel type aortic arch reconstruction, LeCompte manoeuvre, intraventricular rerouting from the left ventricle to the pulmonary arterial valve through a right ventriculotomy using a Dacron® patch and reconstruction of the right ventricular outflow tract using a 12-mm right ventricle to pulmonary artery (RV-PA) PTFE conduit. Her postoperative course was complicated by a prolonged intensive care unit course and a late redo sternotomy with neoaoortic valve repair. Following the second surgery, the patient’s condition stabilized and she was subsequently discharged home. Five months later, the child presented with a shock-like picture and was found to have a severe RV-PA conduit stenosis at the site of the insertion of the conduit into the branch PA’s by cardiac catheterization, and she had minimal response by balloon angioplasty. She later underwent a repeat...
surgery with a 20 mm PTFE RV-PA conduit replacement and repair of branch PAs using a bovine pericardial patch. She was doing well on her recent follow-up.

DISCUSSION

Inclusion of our two patients increases the total reported patients with right-sided IAA in the literature to 25 [1–4]. Right-sided IAA is an extremely rare anomaly with no known true incidence. As reported previously [1], interruptions of right-sided aortic arches are usually type B interruptions except for a single case report in which the patient who was positive for 22q11 chromosomal microdeletion had a right-sided type A IAA with a complete atrio-ventricular canal. That patient underwent a successful surgical repair with direct termino-lateral anastomosis between the ascending and descending aorta, and central pulmonary artery band as a neonate [2].

Both our patients have an aberrant subclavian artery arising from the descending aorta, making a total of 11 patients with an anomalous origin of the subclavian artery historically. McElhinney et al. [1] summarized all the reported right-sided IAA cases to 1999 in which the DiGeorge syndrome was documented in 11 of 16 patients. Of the remaining five patients, subclinical DiGeorge syndrome or del22q11 was suspected in at least some of those patients. Some of these speculations arose from the fact that genetic testing was not routinely available. In recent case reports, all the patients were positive for the DiGeorge syndrome/22q11 chromosomal microdeletion [2–4].

In our findings, Patient 1 was fluorescence in situ hybridization positive for the DiGeorge syndrome, and Patient 2 had a microdeletion of chromosome 22q11. Thus, the total number of confirmed DiGeorge syndrome cases, including our patients is 17 of 25 patients (68%). Neonatal repair of IAAs has become a common practice; there are several reported procedures [4, 5] that included but were not limited to, palliative procedures like stenting the ductus, single stage repair with extended
end-to-end anastomosis with or without subclavian artery ligation, using the aberrant subclavian artery to reconstruct the aortic arch and direct anastomosis by sparing the aberrant subclavian artery. Staged repair using interposition grafts has fallen out of favour due to the improvements in neonatal surgical techniques and the need for follow-up surgeries [5].

For the first time, we report the feasibility of placing an interposition graft from the right chest in a patient with unapproachable descending aorta via median sternotomy, and a modified Yasui procedure with the LeCompte manoeuvre. The modified Yasui procedure is being increasingly used in patients with severe left ventricular outflow tract obstruction (LVOTO) with IAA and is well-described in patients with left-sided IAA, though earlier studies have not shown favourable results with Yasui repair of IAA [5]. Our current approach to IAA is complete primary repair, either by modified Yasui procedure in patients with severe LVOTO or by direct anastomosis with VSD closure. In both our patients, subclavian arteries were preserved, and none of the patients had airway problems.

CONCLUSION

Right-sided IAA is an extremely rare malformation; type B interruptions are common and are often associated with the DiGeorge syndrome/22q11 chromosomal microdeletion. We report two patients with right-sided IAA and the feasibility of innovative surgical repairs.

Conflict of interest: None declared.

REFERENCES