Cardiology in the Young

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Single ventricle palliation of truncus arteriosus with mitral atresia, interrupted aortic arch, and aberrant right subclavian artery

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Brief Report

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Abstract

The association of truncus arteriosus communis with interrupted aortic arch and mitral atresia is an exceptionally rare congenital defect. We describe the initial decision-making and management of this lesion, which eventually achieved a Fontan palliation.

Truncus arteriosus communis is an uncommon form of CHD, which accounts for 1–2% of all congenital heart lesions.[1] Aortic arch interruption occurs in about 10–15% of cases of truncus arteriosus communis, and the preferred management strategy is complete repair during the neonatal period. The association of atrioventricular valve atresia is extremely uncommon and requires palliation towards a Fontan circulation.[1,2] We present a case of truncus arteriosus communis with mitral atresia and hypoplastic left ventricle associated with interrupted aortic arch and aberrant right subclavian artery who underwent Stage I Norwood palliation and subsequently Fontan completion.

Case report

A female with a prenatal diagnosis of hypoplastic left heart and interrupted aortic arch was delivered via C-section at term with a weight of 2.3 kg. Prenatal history included gestational diabetes and polyhydramnios. Apgar scores were 9 and 9, and prostaglandin infusion was initiated.

She was well perfused with peripheral oxygen saturation of 95% and a 13 mmHg blood pressure gradient between the right arm and lower extremities. Cardiovascular exam revealed an III/VI continuous murmur at the base. She also had a talipes equinovarus.

Echocardiogram revealed a hypoplastic left heart with truncus arteriosus communis, an interrupted arch, and a dilated right ventricle with normal systolic function. There was a small muscular VSD with bidirectional shunting. The mitral valve was atretic, and the ascending aorta was hypoplastic. The truncal valve had thickened and doming leaflets with a flow velocity of 2.2 m/second and no regurgitation. There was a large PDA with bidirectional flow, unobstructed left to right flow at the atrial level, and an aberrant right subclavian artery from the descending aorta. A CT angiogram was obtained to further delineate the arch anatomy (Fig 1). Diagnosis of truncus type A4 (Van Praagh) with an aberrant right subclavian, as well as hypoplastic left heart syndrome with mitral atresia, was confirmed, and PGE1 was discontinued. Genetic testing confirmed 22q11 deletion.

On DOL 7, she underwent a stage I Norwood procedure including reconstruction of the interrupted aortic arch. The branch pulmonary arteries were excised from the posterior aspect of the truncus and sutured together to make a confluence. Based on clinical condition and size, the patient received a 5 mm right ventricle to PA conduit, which was revised intraoperatively to 6 mm due to evidence of low PaO_2 despite ventilation with supplemental oxygen and nitric oxide Echocardiogram demonstrated excellent right ventricular function, trivial truncal valve regurgitation, unobstructed flow in both branch pulmonary arteries, and no arch obstruction. She received milrinone and nitric oxide in the ICU.

Delayed sternal closure occurred on post-operative day 4. Following demonstration of left vocal cord paresis and aspiration of liquids by barium swallow, she underwent a gastric tube placement, along with a Nissen fundoplication. She was discharged to home on post-operative day 58.

Catheterisation pre-stage II showed mild truncal valve stenosis (gradient of 12 mmHg), no truncal insufficiency, normal right ventricle function, normal pulmonary resistance, and a low transpulmonary gradient of 4–7 mmHg. Balloon dilatation of a recurrent distal arch stenosis was performed. On DOL 141, she underwent a hemi-Fontan procedure, atrial septectomy, and takedown of the right ventricle to PA conduit.

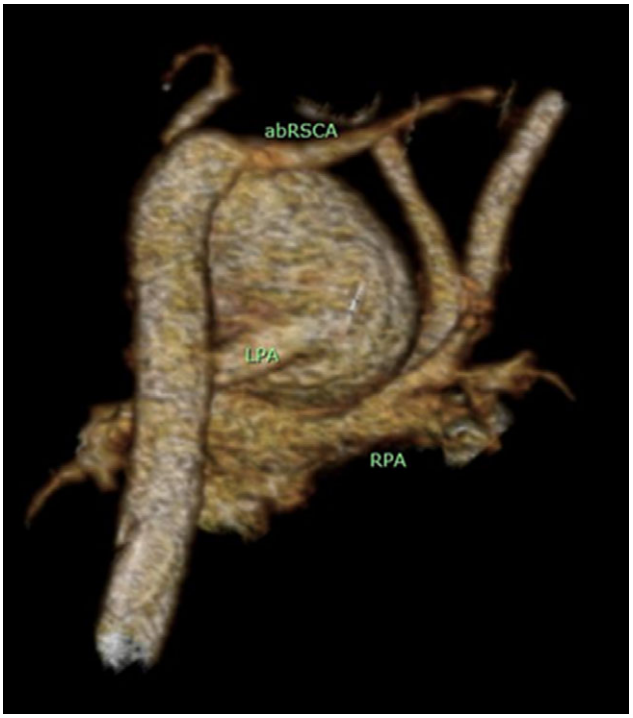

Figure 1. Posterior view of a CT angiogram reconstruction, illustrating the truncus arteriosus, hypoplastic ascending aorta, interrupted aortic arch, and aberrant right subclavian artery. Note that the LPA arose from a very bulbous proximal MPA and somewhat distal and superior to the origin of the RPA. abRCCA = aberrant right subclavian artery; LPA = left pulmonary artery; RPA = right pulmonary artery.

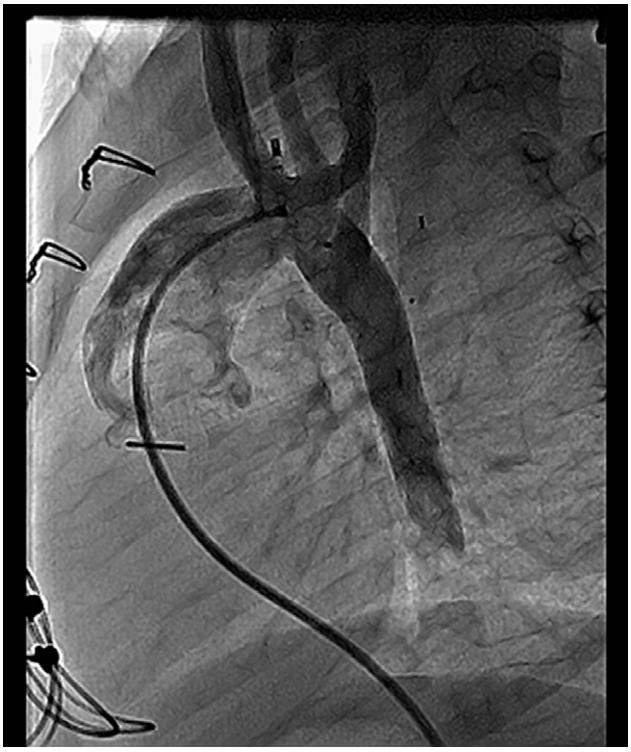

Figure 2. Lateral angiogram of the reconstructed aortic arch.

At 20 months of age, she was noted to have bradycardia with spontaneous second-degree heart block due to which she underwent pacemaker implantation, and subsequently, she had a successful Fontan completion at 3.5 years of age.

Comment

Truncus arteriosus with mitral valve abnormalities has been previously reported, including successful palliation towards a single ventricle.[1–5] The association of truncus arteriosus with interrupted aortic arch and mitral atresia is extremely unusual and not well described. The Congenital Heart Surgeons Society reported the largest experience with truncus and interrupted aortic arch. Among 50 neonates, only 6 had an aberrant subclavian artery, and the only patient with single ventricle physiology was referred for heart transplantation, which confirms the rarity of the constellation of lesions presented and the challenge it represents.[6] While management of patients with single ventricle physiology has been associated with significant improvements and reproducible outcomes, this case required physiologic inferences and technical modifications to optimise the palliation towards a Fontan circulation. The initial perioperative management was similar to hypoplastic left heart syndrome and focused on maintaining patency of the ductus arteriosus and a balance between systemic and pulmonary circulations. A CT was chosen to better delineate the anatomy of the arch and its branches. The initial palliative procedure was a Stage I Norwood, which had the added complexity of repairing the interrupted arch in association with the aberrant right subclavian artery. However, despite the arch interruption, the presence of a common truncus simplified the perfusion, as only one arterial cannula was required. The arch was reconstructed creating a posterior wall of native tissue and augmented with pulmonary homograft (Fig 2), avoiding the use of an interposition graft, therefore decreasing the recurrent need for reintervention described in the CHSS cohort.

Although the truncal valve initially exhibited a mild to moderate gradient on echocardiogram, this was in the presence of a high Qp/Qs. Nevertheless, we chose to provide pulmonary flow using a right ventricle to pulmonary artery conduit in order to decrease total flow through this area. In contradistinction to the report by Michelfelder et al,[4] the proximal pulmonary arteries were mobilised from the truncus, with careful dissection near a commissural post and the left coronary ostium. This facilitated the reconstruction of the pulmonary confluence. Despite sound haemodynamics, cardiac function and balanced cardiac output, the patient had a prolonged hospital course and received a gastrostomy and fundoplication. Moreover, the patient exhibited an elevated pulmonary vascular resistance, which was managed by upsizing the right ventricle to PA conduit to 6 mm plus the addition of oxygen, nitric oxide and sildenafil. Subsequently, a cardiac catheterisation demonstrated normal pulmonary vascular resistance and no conduit or pulmonary branch obstruction. As conditions were favourable, the patient underwent hemi-Fontan followed by Fontan completion. Interestingly, several months after stage II, she exhibited bradycardia associated with second-degree heart block, which could not be explained and for which she received a dual-chamber pacemaker.

We illustrate a very rare case of complex truncus arteriosus with interrupted arch, mitral atresia, and aberrant subclavian artery, which based on physiologic principles and normalisation of cardiac work could be effectively palliated achieving a good functional outcome while avoiding cardiac transplantation.

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Ethical standards. The authors assert that all procedures contributing to this work comply with the ethical standards of the relevant national guidelines on human experimentation (Human subject protection) and with the Helsinki Declaration of 1975, as revised in 2008, and consent has been waived by the institutional review board of the Alfred I DuPont Hospital for Children.

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