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Brief Report

Surgical approach to a rare case of double-outlet right ventricle and aortopulmonary window

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Abstract Although the spectrum of physiology seen in infants and children with double-outlet right ventricle is broad, this anatomy in combination with an aortopulmonary window is extremely rare. We present an interesting case of an infant prenatally diagnosed with this rare CHD. To our knowledge, this is the first report of complete repair in the neonatal period for such a combination of defects.

Keywords: Aortopulmonary window; cardiothoracic surgery; CHD; double-outlet right ventricle

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Case report

An infant was diagnosed in utero with complex CHD including double-outlet right ventricle, a large ventricular septal defect, D-malposition of the great arteries, and a large aortopulmonary window. Given this extremely unusual combination of lesions and the fact that complete neonatal repair has not been previously reported, we describe this patient's cardiovascular anatomy and our approach to surgical management.

The patient was a term male delivered at 38 weeks of gestation via caesarean section. After birth, he remained hemodynamically stable with oxygen saturations ranging from 86 to 95%. A transthoracic echocardiogram demonstrated levocardia, atrial and visceral situs solitus, and D-looped ventricles. Both the aortic and pulmonary outflow tracts arose from the right ventricle and were D-malposed (Supplemental Figure S1). The right atrium was mildly dilated and the left atrium was normal in size. There was a patent foramen ovale with several additional small fenestrations present in the atrial septum. There were no atrioventricular valve abnormalities. A moderate-to-large, unrestrictive, remote ventricular septal defect was present. The left ventricle was normal in size. The right ventricle was mild-to-moderately dilated and was co-apex forming. Biventricular systolic function was normal. The pulmonary valve and the proximal main pulmonary artery were moderately hypoplastic, measuring 3.5 mm (z score: −4.9)[1] and 3.8 mm (z score: −4.6),[1] respectively. There was also sub-valvar narrowing with resultant severe pulmonary stenosis. There was no obstruction to aortic outflow. A large aortopulmonary window was present between the ascending aorta and the distal main pulmonary artery at the junction of the main and right pulmonary arteries (Supplementary Figure S2). The ductus arteriosus was absent. The branch pulmonary arteries were normal in size. The origins of the coronary arteries were anomalous with a common origin of the right coronary artery and left anterior descending coronary artery from the anterior and leftward facing sinus of Valsalva. The circumflex coronary artery arose from the right side of the posterior-facing sinus of Valsalva. The patient remained stable in the ICU and was taken to the operating room on the 7th day of life for complete surgical repair.

Inspection of the patient's external cardiac anatomy in the operating room was consistent with the

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pre-operative imaging findings. The aortopulmonary window connection was located adjacent to the pulmonary artery bifurcation. After the patient was placed on cardiopulmonary bypass and the heart arrested with cold blood cardioplegia, the connection between the aorta and the pulmonary artery was completely transected, and the aorta was closed with a bovine pericardial patch. A vertical right ventriculotomy was made to address the ventricular septal defect. This defect measured ~7 mm and was noted to be a remote defect, but located closer in proximity to the aortic valve than to the pulmonary valve. The ventricular septal defect was enlarged anteriorly and superiorly to a diameter of 10 mm and was baffled to the aortic valve with a Dacron patch. The patient's anomalous left anterior descending coronary artery crossed the right ventricular outflow tract, precluding a transannular patch from being a feasible option. The main pulmonary artery was, therefore, transected and suture ligated. Right ventricular to pulmonary artery continuity was re-established by placement of a 12-mm Contegra® bovine jugular vein conduit (Medtronic, Inc., Minneapolis, Minnesota, United States of America). A transesophageal echocardiogram performed at the conclusion of the operation demonstrated a widely patent right ventricular to pulmonary artery conduit with no obstruction and mild insufficiency, an intact aorta, and a small hemodynamically insignificant residual ventricular septal defect with left-to-right shunting. Biventricular systolic function was normal. His post-operative course was notable for successful resuscitation from a bradycardiac arrest event in the immediate post-operative period, likely secondary to a pulmonary hypertensive crisis. Delayed sternal closure was performed on the 6th day after surgery. The patient underwent elective uncomplicated conduit replacement at 6 months of age for progressive distal conduit stenosis. He was alive and thriving at his last follow-up evaluation at 17 months of age.

Discussion

Double-outlet right ventricle and aortopulmonary window are both rare lesions, each accounting for <1% of all CHDs.[2,3] Although the definition of double-outlet right ventricle has been previously debated,[4] we classified this patient as having such anatomy as both the great arteries were committed to the morphological right ventricle. The specific pathogenesis of the aortopulmonary window remains incompletely understood,[5] but results from a failure of differentiation of the conotruncus into the aorta and pulmonary artery. To our knowledge, no significant clinical data exist about the association between these two defects. Some basic science research suggests that abnormal neural crest cell migration is a possible embryological explanation.[6]

As double-outlet right ventricle and aortopulmonary window are very rarely seen in combination, experience in the management of these patients is limited. A case report by Cabano et al[7] described an infant exposed to maternal phenylketonuria with similar anatomy in whom repair of the aortopulmonary septal defect was accomplished at 1 week of age with subsequent complete repair at 5 months of age. Although this was described as being a successful approach, we favour complete neonatal repair whenever feasible. In addition, such a strategy was not felt possible for our patient, as his degree of pulmonary obstruction warranted intervention at the time of repair of the aortopulmonary window in order to maintain adequate pulmonary blood flow. In a second report, Cavalini et al[8] described a 2-year-old child diagnosed with double-outlet right ventricle, a restrictive remote ventricular septal defect, supravalvar mitral ring, and an aortopulmonary window. At the time of presentation, this patient had already developed pulmonary hypertension and significant left ventricular dysfunction. We believe this report further highlights the importance of early diagnosis and neonatal surgical repair before the onset of significant and potentially irreversible haemodynamic changes.

As our patient required re-operation due to conduit stenosis at 6 months of age, we recognised that an alternative operative strategy could include utilisation of a transannular patch if the anatomy is amenable. Such an approach may reduce the need for early re-intervention; however, the course of our patient's anomalous left anterior descending coronary artery made this approach not feasible.

Double-outlet right ventricle is associated with aneuploidy syndromes including trisomy 13 and trisomy 18, as well as 22q11 deletion.[2] Specific genetic abnormalities in patients with aortopulmonary window remain unknown. Chromosomal microarray obtained in our patient demonstrated interstitial duplication of at least 367 kb of DNA from 5p12 and interstitial deletion of at least 350 kb of DNA from 16p12.2. These findings are of unclear clinical significance.

In conclusion, the combination of double-outlet right ventricle and aortopulmonary window is an extremely rare combination of CHDs. Complete surgical repair in the neonatal period involving direct repair of the aortopulmonary window, ventricular septal defect closure, and right ventricle to pulmonary artery conduit placement can be safely accomplished.

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Conflicts of Interest

None.

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 and with the Helsinki Declaration of 1975, as revised in 2008.

Supplementary material

To view the supplementary material for this article, please visit http://dx.doi.org/10.1017/S1047951115000049

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