Case report

Neonatal repair of right interrupted aortic arch, aberrant left subclavian artery, ventricular septal defect and retroaortic innominate vein

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Abstract

We report a rare case of neonatal biventricular repair of a right interrupted aortic arch (type B), with an aberrant left subclavian artery, ventricular septal defect and retroaortic innominate vein in a 4-week-old, 2.7 kg neonate with DiGeorge syndrome. The patient had an unremarkable postoperative recovery. We discuss the anatomy of this rare congenital anomaly, its surgical implications and issues surrounding the adequacy of the left ventricular outflow tract.

Keywords: Right interrupted aortic arch; Aberrant subclavian artery; Neonatal repair; Biventricular repair; DiGeorge syndrome; Congenital heart disease

1. Introduction

Interrupted aortic arch is commonly associated with a ventricular septal defect (VSD) and almost always a left sided descending aorta (DA) [1]. Right interrupted aortic arch and descending aorta is exceedingly rare [2,3]. Relative unfamiliarity with this presentation raises potential concerns about methods of aortic arch reconstruction. As with left sided interrupted aortic arch and an aberrant right subclavian artery, questions regarding the adequacy of the left ventricular outflow tract (LVOT) are paramount because they affect re-intervention rates and survival following repair [1].

2. Clinical summary

A 4-week-old, 2.7 kg term neonate presented in severe respiratory distress and acidosis, precipitating cardiopulmonary arrest requiring chest compressions. Two-dimensional echocardiography revealed aortic arch interruption beyond the common carotid arteries, with a large outlet VSD, and a retroaortic left innominate vein. The aortic valve was bicuspid with an annular diameter of 3.8 mm. Magnetic resonance (MR) imaging (Figs. 1 and 2) confirmed these findings and identified a right sided ductal arch that continued over the right mainstem bronchus into a right sided descending aorta, with an aberrant left subclavian artery and dense bibasilar lung collapse. The presence of a patent arterial duct (PAD) coursing over the right mainstem bronchus resulted in an impressive 90° rightward rotation of the distal main pulmonary artery. This rotation resulted in a foreshortened right pulmonary artery (RPA) assuming an inferior and somewhat posterior location and an elongated left pulmonary artery (LPA) in a superior and anterior location, particularly evident in Fig. 2. She was resuscitated with a prostaglandin E1 infusion. Chromosomal screening identified that she had a 22q11 microdeletion confirming DiGeorge syndrome.

She underwent urgent operation through a standard median sternotomy approach. Cardiopulmonary bypass was initiated via bicaval venous cannulation and distal ascending aorta (AA) and ductal arch cannulation. The VSD was an outlet defect with almost complete absence of the conal septum. As the patient was cooled to 18°C, the left ventricular outflow tract was assessed through the outlet ventricular septal defect. Because a 5 mm probe could be passed through the aortic valve, a conservative approach was used for the LVOT. The ventricular septal defect was closed with a GoreTex® patch through the pulmonary valve, anchoring the upper end of the suture line into the pulmonary valve annulus, with some sutures above the base of the cusps. At deep hypothermia, the
A ductal cannula was removed and antegrade regional cerebral perfusion was maintained during the arch reconstruction through the ascending aortic cannula. The descending aorta was extensively mobilized and the left aberrant subclavian artery was ligated and divided to facilitate a tension-free anastomosis. All ductal tissue was resected. The descending aorta was easily anastomosed to the posterior aspect of the ascending aorta in an end-to-side fashion. The presence of the retroaortic innominate vein did not impair the performance of the direct aortic anastomosis. The patient was successfully weaned off cardiopulmonary bypass with excellent hemodynamics. Postoperatively, she demonstrated good perfusion with excellent femoral pulses and was rapidly weaned off inotropic support. Her postoperative course was marked with bilateral atelectasis which resolved over time and she was extubated on postoperative day 7. Echocardiography demonstrated good biventricular function, no flow acceleration across the aortic arch repair and a mild (peak 27 mmHg) gradient within the left ventricular outflow tract. She has been well at 3 months follow-up.

### 3. Discussion

Right interrupted aortic arch is a rare presentation with only two other reports cited within the literature [2,3]. A recent multi-institutional Congenital Heart Surgery Society study of 472 cases of interrupted aortic arch did not report a single case of a right-sided descending aorta [1]. The defining feature of right-sided interrupted aortic arch is a patent arterial duct coursing over the right mainstem bronchus and continuing posteriorly with the descending aorta. Because the right mainstem bronchus sits higher than the left and the superior caval vein occupies its normal position, we were concerned about the amount of space available for aortic arch reconstruction as well as possible problems mobilizing the descending thoracic aorta. In addition, the presence of a retroaortic innominate vein and the marked rotation of the branch pulmonary arteries further increased these concerns. To our delight, none of these concerns materialized. We performed a direct tension-free anastomosis without patch augmentation because the right descending aorta easily reached the ascending aorta, particularly after dividing the aberrant left subclavian artery.

Interrupted aortic arch was associated with DiGeorge syndrome in 18% of patients within a large series [1]. When considering our patient and the five other previously reported patients with right interrupted aortic arch [2,3], all six cases of right interrupted aortic arch were associated with DiGeorge syndrome.

Two-dimensional echocardiography was useful in describing most of the anatomy of our patient, however, arch laterality and branching were difficult to establish. Magnetic resonance imaging, performed with a 1.5 T magnet (GE medical system version 11), easily identified the right interrupted aortic arch (type B) with the position of the arterial duct over the right mainstem bronchus and the presence of an aberrant left subclavian artery. Three-dimensional MR images were reconstructed using an Advantage Window station (version 4.1) and a gadolinium enhanced MR angiography protocol. The patient was...
sedated, intubated and mechanically ventilated for the duration of the study.

Left ventricular outflow tract obstruction is a well-recognized problem in interrupted aortic arch, particularly in type B interruption where with an aberrant subclavian artery arises from the descending aorta. Surgical options for LVOT obstruction include a conservative approach, direct outflow tract enlargement procedures or LVOT bypass operations (either biventricular or univentricular). At surgery, the aortic valve annulus was sized at 5 mm. We recently described a formula based upon weight and smallest LVOT diameter where conservative management was acceptable if the LVOT diameter was greater than the neonate’s weight plus 2 mm [4]. Accordingly, we felt that a conservative approach to the LVOT would be successful in our patient. Follow up echocardiography demonstrated a peak instantaneous gradient of 27 mmHg across the left ventricular outflow tract.

Acknowledgment

M.W.C. receives financial support as the recipient of the William T. MacEachern Research Fellowship.

References