Deferred Norwood in the setting of airway compression in double-inlet left ventricle with dextro-transposition of the great arteries

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ABSTRACT

A 4.1 kg male neonate with a diagnosis of double-inlet left ventricle with dextro-transposition of the great arteries was intubated shortly after birth due to respiratory insufficiency. The initial management consisted of a successful Stage I hybrid procedure. Persistent respiratory insufficiency led to cross-sectional imaging and bronchoscopy that demonstrated severe airway compression from a dilated main pulmonary artery. A Norwood procedure with Blalock–Thomas–Taussig shunt was performed at 1 month of age to relieve the airway obstruction. The patient was discharged home on room air at 2 months of age. This case highlights a unique single-ventricle anatomic variant with airway compression, which was successfully managed with deferred Norwood palliation.

Keywords: Airway, congenital heart disease, great vessel anomaly, neonate, univentricular heart

INTRODUCTION

Stage I hybrid palliation, in the form of bilateral pulmonary artery banding with ductal stenting or prostaglandin E1 infusion, is utilized as a management strategy for neonates who are deemed high risk for the initial Norwood procedure. Generally accepted high-risk criteria include low birth weight, multiple congenital anomalies, extreme prematurity, respiratory failure, and pulmonary venous obstruction, among others.[1-3] Subsequent management options include either a deferred Norwood or comprehensive Stage II palliation.

Here, we describe the management of a neonate with double-inlet left ventricle and dextro-transposition of the great arteries with unrecognized airway obstruction due to great vessel anatomy who was initially managed with hybrid palliation due to the uncertain etiology of respiratory failure at birth.

CASE REPORT

A 4.1 kg male with a prenatal diagnosis of double-inlet left ventricle with dextro-transposition of the great arteries was emergently delivered through a cesarean section at 36 weeks due to fetal distress. The APGAR scores were 3 and 5 at 5 and 10 min, respectively. He had normal renal and head ultrasounds as well as a normal congenital chromosomal analysis and microarray. He was intubated shortly after birth due to respiratory insufficiency. On initial echocardiography, he was shown to have a double-inlet left ventricle, dextro-transposition of the great arteries (S, D, and D), hypoplastic right ventricle

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with an unrestrictive muscular ventricular septal defect, and a hypoplastic left-sided aortic arch with a large patent ductus arteriosus.

He developed signs of pulmonary overcirculation by the day of life 9, so Stage I hybrid palliation, in the form of bilateral pulmonary artery bands and prostaglandin infusion, was performed on the day of life 11. Cross-sectional imaging and bronchoscopy were performed on postoperative days 17 and 19, respectively, due to persistent respiratory failure requiring invasive positive pressure respiratory support. These demonstrated severe pulsatile compression of the distal trachea and left mainstem bronchus from a dilated main pulmonary artery [Figure 1]. A Norwood procedure with a Blalock–Thomas–Taussig shunt was then performed at 4 weeks of life (postoperative day 21 from Stage I hybrid). The patient was extubated on postoperative day 10 from this procedure and discharged home on room air at 2 months of age. Repeat cross-sectional imaging at 6 months of age demonstrated that the distal trachea and left mainstem bronchus were no longer compressed by the Damus–Kaye–Stansel anastomosis [Figure 2]. He subsequently underwent a successful bidirectional Glenn procedure and is doing well as an outpatient.

DISCUSSION

Double-inlet left ventricle is a rare single ventricle variant that occurs in up to 0.01 per 10,000 live births.[4] Within this subset of patients, up to 16% may present with a rightward right ventricle and discordant ventriculoarterial connection.[5] This patient's anatomy also included a dilated, posteriorly located pulmonary artery that resulted in airway compression, the combination of which in the setting of double-inlet left ventricle and dextro-transposition of the great arteries has been rarely reported. This went unrecognized as the likely cause of respiratory decompensation at the time of birth, and Stage I hybrid was elected as the initial palliative strategy for the patient due to the persistent need for mechanical ventilatory support. Cross-sectional imaging was not obtained until the patient failed attempts at extubation following the initial Stage I hybrid. This clearly demonstrated the airway compression, and we subsequently proceeded with a Norwood procedure at 1 month of age. Transection of the pulmonary artery and creation of the Damus–Kaye–Stansel anastomosis relieved the airway compression sufficiently to allow extubation on the postoperative day 10 from the Norwood.

In retrospect, earlier cross-sectional imaging and/or bronchoscopy may have demonstrated the patient's airway anatomy as the potential cause of respiratory failure. Subsequent bilateral pulmonary artery banding may have contributed to further dilation of the pulmonary artery. In the absence of common high-risk characteristics, we may have elected to proceed with a Norwood as the initial procedure, thus potentially avoiding multiple interventions and a prolonged hospital stay. Nonetheless, our experience demonstrates that airway compression by a dilated pulmonary artery can be a rare but reversible cause of respiratory distress in this anatomy. Furthermore, hybrid palliation of univentricular neonates remains a reasonable management strategy for patients with an unclear risk profile, permitting additional time for further testing while maintaining the option of a deferred Norwood for appropriate candidates.[1]

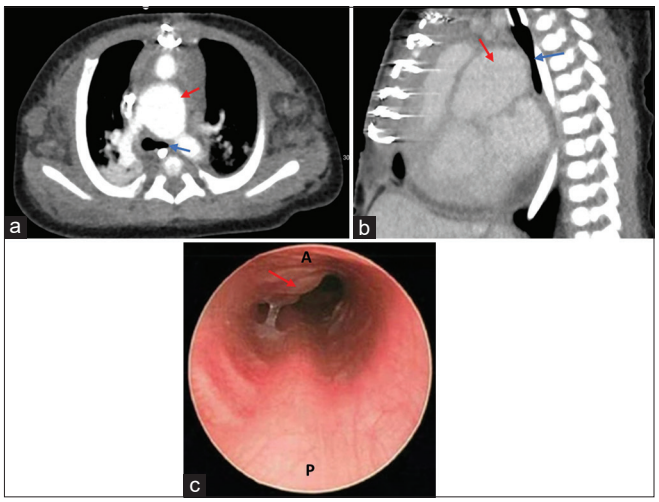

Figure 1: (a) Axial and (b) Sagittal computed tomography imaging showing the dilated main pulmonary artery (red arrow) compressing the left mainstem bronchus (blue arrow) near the carina, (c) Bronchoscopy showing external pulsatile compression from the pulmonary artery (arrowhead) of the distal trachea and left mainstem bronchus

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to

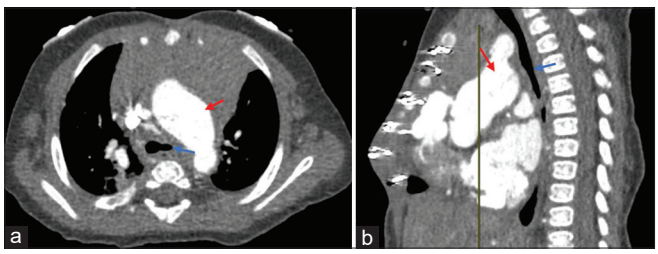

Figure 2: (a) Axial and (b) Sagittal computed tomography imaging showing that the left mainstem bronchus near the carina (blue arrow) is no longer externally compressed by the nearby Damus–Kaye–Stansel anastomosis (red arrow)

be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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