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CASE REPORT

Successful Staged Surgical Repair Using Rapid Pulmonary Artery Banding in a Very-Low-Birth-Weight Premature Infant Who Had d-Transposition of the Great Arteries With an Intact Ventricular Septum

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Abstract The arterial switch operation is the surgical correction of choice for patients born with d-transposition of the great arteries (d-TGA) and an intact ventricular septum. However, prematurity and very low birth weight present both technical and physiologic challenges to this approach. Furthermore, in the setting of d-TGA and an intact ventricular septum, delaying intervention results in deconditioning of the left ventricle, rendering the patient a poor candidate for the arterial switch operation. The report presents an infant born at 27 weeks gestation weighing 1.01 kg who as a newborn underwent a successful urgent balloon atrial septostomy, pulmonary artery banding, and a central shunt on day of life (DOL) 82 and the arterial switch operation on DOL 93.

Keywords Arterial switch operation · d-TGA · d-Transposition of the great arteries · Intact ventricular septum · Pulmonary artery banding

Case Report

A premature baby boy born at 27 weeks gestation with a reported birth weight of 1,010 g was severely hypoxemic and acidotic after birth. After intubation and mechanical ventilation, an echocardiogram showed d-transposition of the great arteries (d-TGA) and restrictive shunting across the foramen ovale.

Prostaglandin E (PGE1) infusion at 0.05 μg/kg/min and dopamine at 5 μg/kg/min were started, and the boy was taken to the cardiac catheterization lab at 6 h of age. He underwent successful balloon atrial septostomy via the umbilical vein approach. At the time of the balloon septostomy, his weight was 990 g, making him one of the smallest reported patients to undergo successful balloon atrial septostomy [15].

The balloon septostomy procedure was performed using the previously described technique [11] but with only partial inflation of a 5-Fr Fogarty septostomy catheter because the small cardiac chamber dimensions made full inflation of the septostomy balloon impossible. The balloon septostomy resulted in a large atrial septal communication, adequate atrial level mixing, and prompt improvement of oxygenation. No vascular compromise was noted after the procedure.

Dopamine infusion was discontinued on DOL 4, and PGE1 infusion was discontinued on DOL 6. The baby was weaned off mechanical ventilation and extubated to continuous positive airway pressure on DOL 6 without complications. After 3 days, the boy was receiving varying fractional inspired oxygen (0.23–0.28) at 2 l/min via a nasal cannula. His saturations were 65–80 %. He required infrequent red blood cell transfusions to maintain a hemoglobin level of 15–18 g/dl.

In light of the patient's extreme prematurity, very low birth weight (VLBW), and the associated increased surgical risk of primary complete repair in his cohort, it was decided to allow for passive left ventricular (LV) "deconditioning" with plans for subsequent "rapid pulmonary artery banding" followed by the arterial switch operation after achievement of adequate somatic growth [5, 12].

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The baby remained in the intensive care nursery, and his diet was progressed, resulting in adequate growth. The boy did not require repeat cardiac catheterization during his hospital stay. Serial clinical and echocardiographic examination showed that mild to moderate dynamic obstruction across the subpulmonary LV outflow tract had developed due to right-to-left septal shift.

On DOL 82, with a weight of 2.7 kg, the boy underwent placement of a pulmonary arterial band and a 3-mm right modified Blalock-Taussig shunt. Every 3 days, he was followed up with serial echocardiograms, which showed increasing LV mass and good LV systolic function. After 11 days, on DOL 93 (corrected gestational age, 39 weeks), with a weight of 3.1 kg, the boy underwent redo sternotomy for takedown of the pulmonary artery band, takedown of the right modified Blalock-Taussig shunt, and the arterial switch operation with LeCompte maneuver, autologous pericardial patch closure of a large atrial septal communication, and autologous pericardial patch augmentation of the neopulmonary root.

The procedure and postoperative recovery were uncomplicated. The boy was discharged home 10 days after the arterial switch operation, on DOL 103, at a weight of 2.94 kg.

At this writing, the boy is 22 months of age and clinically well with very good hemodynamic balance and normal neurodevelopmental status. His echocardiogram at 18 months of age showed no residual atrial communication, widely patent great vessel anastomotic sites with only mild turbulence of antegrade flow across the pulmonary anastomosis (mean Doppler gradient, <10 mmHg), mild neopulmonary valve insufficiency, and mild neoaortic valve insufficiency. He has normal LV mass and normal biventricular systolic function.

Discussion

Among newborns, d-TGA is a common congenital heart defect, representing up to 7 % of all congenital cardiac malformations. In the newborn period, d-TGA, the second most common congenital heart defect, presents with cyanosis. The arterial switch procedure with LeCompte maneuver is the established preferred surgical repair for infants with d-TGA. This procedure is associated with a low operative mortality rate and excellent long-term survival [2, 3, 9]. Before surgical repair, most d-TGA patients, without adequate mixing of oxygenated and deoxygenated blood, require a balloon atrial septostomy [11].

The arterial switch procedure usually is performed during the first 3 weeks of life, before the expected decrease in pulmonary vascular resistance and the consequent drop in LV peak pressure, which in turn causes loss of LV mass and ventricular "deconditioning" [6]. Therapeutic strategies for the premature VLBW patient vary, and the clinical data supporting such strategies are mixed. Some centers advocate early definitive repair independent of gestational age and weight, whereas others defer intervention until a certain gestational age or weight has been achieved.

Prematurity and a VLBW can have a negative impact on outcome in the first few weeks of life due to increased surgical risk, increased risk of cerebral hemorrhage, and the influence of coexisting noncardiac conditions. In patients for whom early surgery is contraindicated or patients presenting after the LV has been deconditioned, a "rapid, two-stage" arterial switch using pulmonary artery banding with or without a shunt for 7–14 days as the initial procedure followed by an arterial switch [1, 10] is an alternative surgical strategy. The initial pulmonary banding results in increased LV afterload, thereby inducing an increase in LV mass [1]. The increased mass of the LV allows for increased ventricular workload capacity and thus a successful surgical outcome after the delayed arterial switch procedure.

The two-stage procedure is associated with a higher morbidity and mortality risk than the neonatal single-stage approach [4, 7, 13, 14]. Some centers have reported success using the late single-stage arterial switch procedure and postoperative mechanical support of cardiac output with extracorporeal membrane oxygenation or a ventricular assist device. Such mechanical support, however, is very difficult in VLBW premature newborns [7, 8].

The reported patient underwent a successful balloon atrial septostomy at 990 g, making him the smallest reported patient to undergo such a procedure via the umbilical vein approach. Moreover, with a birth weight of 1,010 g, he is one of the smallest reported patients to undergo successful two-stage repair of d-TGA. After planned staged surgical repair of d-TGA with an intact interventricular septum, our patient at this writing remains clinically stable with no significant residual cardiac compromise at almost 2 years of age.

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