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Commentary: The end justifies the means: Employing a "modified" strategy for Starnes palliation in a premature low birth weight infant with Ebstein anomaly

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CENTRAL MESSAGE

Management of neonatal Ebstein Anomaly with a circular shunt may require expeditious and novel surgical interventions that comply with but do not necessarily follow standard algorithmic approaches.

Deng and colleagues[1] from The Hospital for Sick Children describe their management strategy for a 32-week gestational age, 1.5-kg newborn with Ebstein anomaly (EA). Interventions included immediate postnatal ligation of the main pulmonary artery (MPA) with right atrial reduction followed by a "modified" Starnes Procedure at 20 hours of life. The group should not only be congratulated for a successful outcome but also for outlining a course in which anatomic approaches in a nontraditional order achieved a desired physiologic state. In additional, multidisciplinary institutional support to provide care for such a high-risk patient deserves recognition.

Several groups have described concerning outcomes for children requiring neonatal intervention for EA and well-accepted algorithmic approaches for their initial care.[2,3] In particular, issues surrounding antegrade pulmonary blood flow and concerns for a circular shunt often direct early treatment strategies. These algorithmic approaches employ mechanical ventilation, inhaled nitric oxide, and infusion of prostaglandin E1 followed by test occlusion of the patent ductus arteriosus as initial strategies. Continued evidence of a circular shunt leads to surgical intervention with Starnes palliation. This process may require several days of support and lead to end-organ compromise.

The group from Sick Kids employed immediate postnatal ligation of the MPA and right atrial reduction to aggressively treat the patient's circular shunt. Due to low cardiac output state, they proceeded with a Starnes procedure and right ventricle exclusion. Due to the patient's size of 1.5 kg, they elected to maintain ductal patency and secure bilateral pulmonary artery bands rather than place a modified Blalock–Taussig shunt. After separation from postcardiotomy extracorporeal membrane oxygenation and recovery, the patient received a Blalock–Taussig shunt at 3 months of age followed by a bidirectional cavopulmonary shunt at 9 months.

The need to eliminate a circular shunt, provide a stable and balanced source of pulmonary blood flow, and allow for adequate cardiac output by decompressing the right ventricle are tenets of surgical care for EA. Immediate MPA ligation followed by a modified Starnes and "hybrid" approach toward balanced pulmonary blood flow allowed this group to successfully achieve these physiologic goals in a premature 1.5-kg neonate. The maintenance of a ductal source of pulmonary blood flow with placement of PA bands is now evolving as an initial treatment strategy for lesions other than hypoplastic left heart syndrome.[4] In addition, the need for postcardiotomy extracorporeal membrane oxygenation and peritoneal dialysis, although only briefly mentioned in the manuscript, required a multidisciplinary institutional focus and support that is often

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challenging, even in the very best of congenital heart centers.

Greater-risk surgical treatment strategies for congenital heart disease are increasingly prevalent. Low birth weight (2.5 kg) as a surrogate for mortality continues to be redefined.[5] A recognition that accepted algorithmic approaches to challenging lesions may require novel and expeditious surgical strategies is an important approach to achieving successful outcomes. Although not involving any degree of "morality," the group from Sick Kids should be congratulated for employing a Machiavellian strategy of being creative in defining the means required to achieve a successful end.

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