Challenges and Controversies in Fetal Diagnosis and Treatment

Hypoplastic Left Heart Syndrome

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KEYWORDS

• Hypoplastic left heart syndrome • Fetal echocardiography
• Fetal aortic valvuloplasty • Norwood operation • Maternal hyperoxygenation

KEY POINTS

- Less than 2 decades ago, hypoplastic left heart syndrome (HLHS) was considered a lethal condition, with most babies dying within days of diagnosis.
- Pediatric cardiologists must be keenly aware of the flaws of staged palliation for the treatment of HLHS.
- Pediatric cardiologists should monitor the emerging data regarding fetal diagnosis and treatment.

INTRODUCTION

During the past 20 years, perhaps no form of congenital heart disease has generated more challenges and controversies than hypoplastic left heart syndrome (HLHS). The surgical approach, initially conceived by William Norwood in the late 1970s, was life-saving, but surgical mortality was high.[1] Rather than abandon the procedure, many centers adopted novel approaches in the management of these patients, resulting in dramatically improved survival (**Fig. 1**).[2–10] Although there is virtually no attrition following the second stage surgical procedure (bidirectional Glenn), the longer-term issues related to Fontan physiology are becoming much more apparent. Morbidities in children with HLHS status post Fontan palliation include exercise intolerance, ventricular dysfunction, progressive tricuspid and neoaortic valve insufficiency, arrhythmias, thromboembolic events, protein-losing enteropathy, plastic bronchitis and hepatic dysfunction, even hepatic carcinoma.[11–23] Of even greater concern is the identification of neurocognitive difficulties in many of these patients, the etiology of

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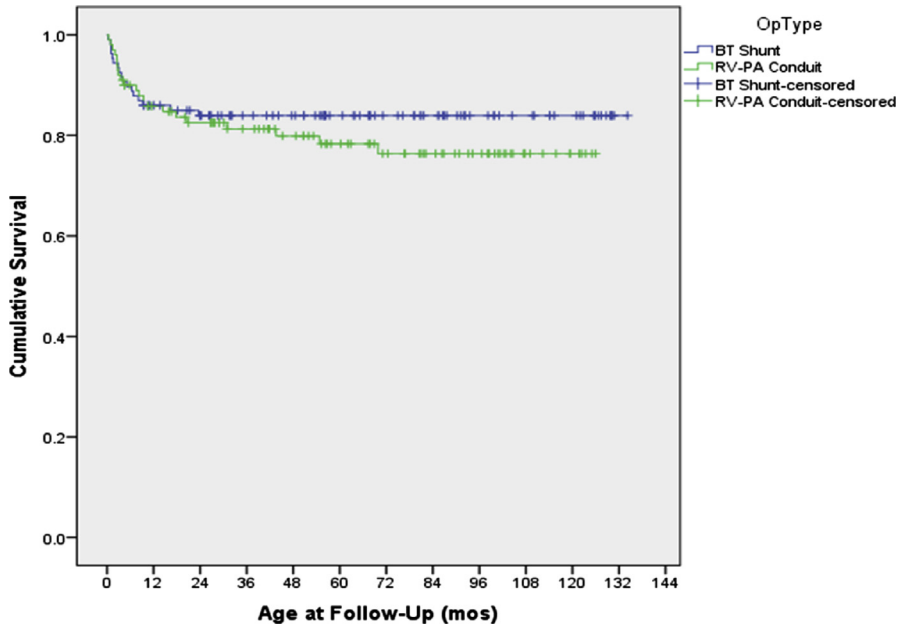

Fig. 1. Actuarial survival, Norwood operation. Children's Hospital of Wisconsin, 2003 to 2013. BT, Blalock-Taussig; RV-PA, right ventricle to pulmonary artery.

which is likely multifactorial.[24–26] Although medical therapy and ventricular assist devices can temper the situation, eventual cardiac replacement therapy for many of these patients seems inevitable. Unfortunately, the risk associated with cardiac transplantation in the Fontan population is high, as patients are frequently listed with significant chronic heart failure symptoms and end organ dysfunction, and there is no consensus on optimal timing of listing.[27–29] Pediatric cardiologists should be keenly aware of the flaws of staged palliation for treatment of HLHS, and need to monitor the emerging data regarding fetal diagnosis and treatment.

ETIOLOGY OF HYPOPLASTIC LEFT HEART SYNDROME

Before the advent of fetal echocardiography, the embryologic cause of HLHS was not entirely clear. However, in 1989 Allan and colleagues[30] observed the in utero evolution of HLHS in a fetus initially diagnosed with critical aortic stenosis. It is now postulated that many cases of HLHS are dynamic and progressive throughout gestation, resulting from altered left ventricular outflow as already described or, less commonly, altered left ventricular inflow (ie, mitral valve stenosis/foramen ovale restriction).[31–33]

In normal fetal circulation, the fetal left ventricle is predominantly filled with oxygenated blood that returns from the placenta and traverses the foramen ovale. If blood flow across the foramen ovale is diminished or reversed, the combined cardiac output is redistributed to the right ventricle and pulmonary artery, resulting in enlargement of the right heart structures and creating less impetus for normal growth of left heart structures. Perhaps the most well-recognized mechanism for decreased flow or reversal of flow through the foramen ovale in utero is the presence of severe aortic valve disease.[34] With significant aortic valve stenosis, alterations in left ventricular compliance occur, either secondary to the development of left ventricular hypertrophy