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Commentary: Will the next improvement in the care of single ventricle patients come from focusing on patient selection for the cavopulmonary shunt?

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CENTRAL MESSAGE

The next steps in the improvement of outcomes in single ventricle patients may come from improved patient selection for the cavopulmonary shunt.

To paraphrase Winston Churchill, although the cavopulmonary shunt (CPS) has issues, it is the best option we have. In 1977, Choussat and colleagues delineated the renowned 10 selection criteria for the Fontan procedure. These guidelines have been revised over the years, with improvements in the assessment and management of patients with single ventricle physiology.¹ More recently, advances have concentrated on early infant care before the CPS.² The period between CPS and Fontan has been relatively overlooked, however. Despite the beneficial transition to a more stable physiology, the CPS is not risk-free. In this issue of the *Journal*, Callahan and colleagues³ address this interstage period and analyze risk factors associated with death/transplantation versus Fontan completion. To minimize confounding factors, they studied patients with tricuspid atresia, a lesion considered to have the most favorable anatomy and outcomes among single ventricle diseases.

Even with the advantageous anatomy of tricuspid atresia, 5% of patients died or underwent transplantation after CPS. Unsurprisingly, moderate or greater mitral valve regurgitation was a risk factor for poor outcome, in line with previous findings.⁴ The study also addressed the controversial issue

of antegrade pulmonary blood flow, the risks and benefits of which have been debated in the literature for more than 2 decades. Some scholars have advocated leaving antegrade flow to the pulmonary arteries to improve pulmonary artery growth and limit arteriovenous malformations. The technique is also used to offset for marginal preoperative anatomy and physiology. Whether the increased risk of death or transplantation with antegrade pulmonary blood flow reflects the preoperative anatomy or the postoperative physiology, the findings of Callahan and colleagues support those of a recent study by Baek and colleagues,⁵ which also demonstrated a survival disadvantage.

Technical factors also influence results. Postoperative superior vena cava intervention was associated with death or transplantation. Early identification and management of residual lesions has become a tenet for improving outcomes.⁶ Recognizing issues with the CPS can be challenging with traditional intraoperative transesophageal echocardiography.⁷ Regardless, the conclusions reinforce that intraoperative assessment of the cavopulmonary circuit to identify and immediately address residual lesions can improve outcomes.

Is it time to start thinking about criteria similar to Choussat's Fontan commandments for the CPS? We all know that some CPS patients do not do well. Improvements in surgical outcomes are often a result of refinements in patient selection. Identifying high-risk patients will aid preoperative planning and development of alternate or staged care strategies. The field has developed

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beneficial detailed approaches to the care of single ventricle patients. The next steps for improvement may well come from the second interstage.

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Commentary: There are no facts, only interpretations

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In the current issue of the *Journal*, Callahan and colleagues¹ report their study of infants born with tricuspid atresia and normally related great arteries from the Congenital Heart Surgeons Society (CHSS) cohort. They focused on outcomes following cavopulmonary shunt (CPS) and examined factors associated with Fontan completion, death, or transplantation following CPS. They found that by 5 years after CPS, 5% of the patients died or underwent transplantation and 91% transitioned to Fontan. They determined that preoperative moderate or greater mitral regurgitation, concomitant mitral repair, a pulmonary artery band (PAB) at CPS, and postoperative superior vena cava interventions, including CPS takedown, were associated with death or transplantation. In addition, a PAB at CPS and prenatal diagnosis were associated with decreased Fontan completion. They concluded that several risk factors contributed

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CENTRAL MESSAGE

Causality between risk factors and outcomes might be difficult to explain, thus limiting our ability to recommend practice changes that would improve results.

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to the mortality following CPS, and that a PAB at CPS contributed to both death and lack of progression toward Fontan.¹

The CHSS has accumulated a large number of patients who were enrolled in numerous study cohorts, including patients with tricuspid atresia. Their sophisticated analyses of these cohorts have contributed to our understanding and subsequently have helped shape the management strategies for various complex congenital heart defects. Advantages of these CHSS studies include the large cohort, the comprehensive dataset, and the long follow-up. Disadvantages include the differing practice patterns among centers and the common lack of clinical information that often