Commentary: Double-outlet right ventricle—Toward an individualized road map

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The optimal definition, classification, and management of double-outlet right ventricle (DORV) are still a matter of debate.¹ Part of the difficulty stems from the wide anatomic and physiologic heterogeneity that characterizes this disease. This same heterogeneity makes studying DORV particularly challenging.

In this issue of the *Journal*, Oladunjoye and colleagues² present a large retrospective series of patients with DORV who underwent biventricular repair. They compared patients who underwent primary repair (including those with a previous isolated pulmonary band) with patients who underwent staged repair (patients who had other palliative procedures, such as shunts or other intracardiac operations, before final repair). Oladunjoye and colleagues² concluded that patients with complex DORV may benefit from a staged repair, because the outcomes of patients with a staged repair were comparable with those of patients with a primary repair, who presumably represented a less complex group of patients. One of the obvious limitations of the study was that patients in both groups were not the same; however, the fact that they were able to achieve relatively similar outcomes despite the higher complexity is an important conclusion in and of itself.

The morbidity and mortality of DORV repair are still significant. In this study, it was estimated that 9% to 16% of patients (depending on the group) would die, and 35% would require surgical reintervention within 3 years. These results are a humbling reminder that surgical “repair” of DORV is best described as a palliative procedure. And just like other palliative procedures in congenital heart surgery, each surgical intervention should not be seen as a goal in itself but rather as a step in a carefully orchestrated long-term surgical “road map” with the destination of providing the patient with the best quantity and quality of life.

A particularly challenging group of patients with DORV are those with a noncommitted VSD, which made up the

Three-dimensional computed tomography of complex double-outlet right ventricle.

Central Message

Double-outlet right ventricle is a complex and heterogenous diagnosis, requiring individualized care by a multidisciplinary team, including advanced tools for anatomic and surgical planning.

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FIGURE 1. Three-dimensional computed tomographic reconstruction of a patient with double-outlet right ventricle with large atrial and septal defects, as well as supracardiac total anomalous pulmonary venous return. The patient underwent imaging after a left sided modified Blalock-Taussig shunt. ©2019 University of Texas Dell Medical School, reprinted with permission.

bulk of patients in the staged cohort of Oladunjoye and colleagues.² The long-term morbidity of a complex biventricular “repair” in these patients is not insignificant. Approximately 50% of them required a surgical reintervention within 3 years, and almost 20% required a permanent pacemaker. Would any of these patients have benefited from single-ventricle palliation instead?³ This study does not allow us to answer this question, because median follow-up for staged repair patients was only 12 months.² For us to answer this question, we will need studies with much longer follow-up and studies that address the long-term psychosocial burden of living with the disease, studies that have been conspicuously missing in our field.

Despite its inherent limitations, this study² provides additional data to help us make decisions in managing this challenging subset of patients. We agree with Oladunjoye and colleagues² that a staged approach to patients with complex DORV is generally advisable, but treatment needs to be

individually tailored to each patient. The use of 3-dimensional imaging and printing tools (Figure 1) will undoubtedly help with the creation of a customized patient road map that may include more than one intervention to arrive at the ultimate destination: the best long-term anatomic, physiologic, and psychosocial outcome for a particular patient.

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