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Commentary: Unwanted baggage tossed or carried on the single ventricle journey?

David P. Bichell, MD

In this issue of the *Journal*, Wilder and Caldarone¹ give a guided tour of the hybrid versus Norwood conversation, noting that bias in patient selection poisons equipoise when trying to compare the merits of the 2 strategies. The authors lay out a thoughtful framework for when and why to apply the hybrid approach to high-risk patients. Most centers report similar outcomes for either strategy and reserve the hybrid for special high-risk cases. Defining the niche for the hybrid approach is an incomplete story that will require defining subcategories of high risk that are modifiable or nonmodifiable, with eyes on the long game.

Operative survival through first- or second-stage palliations is not the same as long-term success. Some risks are carried as baggage to the second stage, third stage, or even beyond transplantation. Tanem and colleagues² found that patients with high pre-stage 1 risk accounts for 83% of pre-Glenn deaths and 63% of deaths before 1 year. Transplantation-free survival was 89% for standard-risk patients versus 54% for high-risk patients. High-risk status does not resolve after Glenn, as long-term survival continues to diverge from the standard-risk group.² Choosing the best method of getting to stage 2 is no assurance that intrinsic risk has been leveled.

Primary transplantation for high-risk patients is not a risk neutralizer either. After accounting for waiting list attrition and post-transplantation mortality, 5-year survival is 54% for primary transplantation for hypoplastic left heart

OUTCOMES ACCORDING TO PREOPERATIVE RISK			
	STANDARD RISK	SOME RISKS	SOME OTHER RISKS
NORWOOD	GOOD	BAD	BAD
HYBRID	GOOD	BAD	MAYBE BETTER

It remains unclear which preoperative risks can be modified by a hybrid approach.

CENTRAL MESSAGE

Determining what advantage can come from the selective use of the hybrid stage 1 for high-risk patients results in a better understanding of which risks are modifiable beyond the short term.

syndrome and 53% for post-Norwood transplantation.^{3,4} So the high-risk patients approach coin toss odds for long-term survival whether palliated, primarily transplanted, or palliated then transplanted, and maybe irrespective of the approach at stage 1.

Evidence suggests that some high-risk features may not be modifiable regardless of strategy. For low birth weight infants, a strategy of delaying surgery until weight gain might not mitigate the risk of mortality at cardiac surgery.⁵ In utero left atrial hypertension from atrial restriction promotes pulmonary arteriolar and venous thickening, lymphatic dilation, and high pulmonary resistance that may persist to haunt the success of the Glenn, Fontan, or transplantation procedure.^{6,7} Although valvuloplasty is successful in reducing regurgitation in the majority of patients with hypoplastic left heart syndrome with the risk factor of tricuspid insufficiency, outcomes are restricted by limited repair durability, with recurrent significant regurgitation in one-third of the patients. Right ventricular dysfunction in these patients is progressive and a major determinant of transplantation-free survival.⁸

Which risks can be favorably modified by a hybrid approach over Norwood? Some of the features that define high risk cannot be neutralized by any stage 1 strategy. The equation to determine who may benefit from a hybrid versus a Norwood approach is not about the inherent superiority of one strategy over another to achieve success at stage 1, but rather is about figuring out which risks a hybrid might modify with durable benefit. Obfuscating the job of defining who might benefit from hybrid are

From the Department of Cardiac Surgery, Monroe Carell Jr Children’s Hospital, Vanderbilt University Medical Center, Nashville, Tenn.

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Address for reprints: David P. Bichell, MD, Pediatric Cardiac Surgery, William S. Stoney Jr Chair in Cardiac Surgery, Monroe Carell Jr Children’s Hospital, Vanderbilt University Medical Center, 5247 Doctors’ Office Tower, 2200 Children’s Way, Nashville, TN 37232-9292 (E-mail: david.bichell@vumc.org).

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some risks that persist despite early survival. Wilder and Calderone bring some guiding clarity to appropriate application of the hybrid. A more in-depth dissection of risk is needed.

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