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Commentary: Comparison of management strategies for the neonate with symptomatic tetralogy of Fallot and weight <2.5 kg—jogging towards equipoise?

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For the newborn with tetralogy of Fallot (TOF) whose cyanosis mandates intervention in the neonatal period, caregivers choose between 2 options: primary repair (PR) or initial palliation (IP) followed by delayed complete repair (CR). Which is better, and what outcomes should be used to decide?^{1,2} Hypothetical arguments in favor of PR include the earliest achievement of “normal” physiology, prevention of morbidity attendant to palliation, and avoidance of a second invasive intervention in the first year of life. Considerations in favor of staging include avoidance of cardiopulmonary bypass (or deep hypothermic circulatory arrest) in the vulnerable newborn period, with a consequent reduction in morbidity and mortality, and performance of the repair when it is simpler (in a larger child), with a consequent reduction in early reintervention. To date, there have been no prospective trials to guide the choice of strategy, and much of the literature has not stratified TOF patients by size, pulmonary artery architecture, pulmonary valve patency, or other important risk factors. Against this backdrop, Qureshi and colleagues from the Congenital Cardiac Research Collaborative report a retrospective comparison of small (<2.5 kg) neonates with cyanotic TOF undergoing IP/CR (n = 76) or CR (n = 44).³ At the IP stage, 23 neonates underwent a transcatheter approach (right ventricular outflow tract [RVOT] stent, patent ductus arteriosus stent, or

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CENTRAL MESSAGE

For cyanotic neonates weighing <2.5 kg with tetralogy of Fallot, neonatal repair and neonatal palliation/delayed repair result in similar survival and reintervention rates at a 5-year follow-up.

balloon pulmonary valvuloplasty), and 53 underwent surgical palliation (RVOT procedure or systemic to pulmonary artery shunt). Very importantly, the authors included not only periprocedural morbidity and mortality, but also continued the period of observation out to several years to allow capture of later reinterventions and late mortality. With median follow-up of 5.5 years, the authors showed similar overall mortality and postrepair reintervention rates for IP/CR and PR. The adjusted procedural and neonatal morbidity burden was lower in the staged repair group, but cumulative secondary outcomes favored the primary repair group.

A striking outcome of this study is that the overall mortality of these patients, irrespective of treatment strategy, was very high at 16.7%, a rate 6% to 11% higher than reported in a companion study by the same authors in cyanotic TOF neonates of normal birth weight.⁴ This excess mortality risk for small patients is similar in magnitude to that recently reported across a spectrum of congenital cardiac anomalies.⁵ Perhaps the more important finding of this study is the fact that neither approach is inarguably superior, and so we will continue to argue. “Stagers” will emphasize the ever-improving results with transcatheter palliation and suggest that the present work is of only historical relevance. “Primaries” will emphasize the lower cumulative morbidity burden (at least in the first year) of a single intervention. As a more constructive alternative, we might instead consider the findings most useful in supporting equipoise. This in

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Disclosures: The authors reported no conflicts of interest.

The Journal policy requires editors and reviewers to disclose conflicts of interest and to decline handling or reviewing manuscripts for which they may have a conflict of interest. The editors and reviewers of this article have no conflicts of interest.

Received for publication Feb 10, 2021; revisions received Feb 10, 2021; accepted for publication Feb 11, 2021; available ahead of print Feb 19, 2021.

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J Thorac Cardiovasc Surg 2022;163:208-9

0022-5223/\$36.00

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<https://doi.org/10.1016/j.jtcvs.2021.02.040>

turn may finally allow for a prospective study with appropriate stratification by birth weight and other important factors. Then we will doubtless find something else to argue about. In the meanwhile, it is clear that both stagers and primaries have work to do to provide more effective and less morbid treatments for our tiniest patients.

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Commentary: Very nice study...what are we to do with it?

Charles D. Fraser, Jr, MD

Another paper on tetralogy of Fallot (TOF)...really? Cardiologists and surgeons responsible for treating children with TOF are confident and entrenched in their opinions about strategy. We counsel with confidence, almost at times being dismissive about the complexity of TOF. It's a straightforward diagnosis and we know what to do. Or do we?

This is a problematic paper, like a road to nowhere. Qureshi and colleagues¹ expose problems we probably weren't expecting. In babies (<2.5 kg) with symptomatic TOF treated with 2 opposing strategies, initial palliation (surgical or catheter-based) followed by a complete repair or by a primary surgical repair without preceding palliation, results aren't great.

Charles D. Fraser, Jr, MD

CENTRAL MESSAGE

This is yet another tetralogy of Fallot management study... or is it? The question is what are we to do to improve outcomes in this disease?

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Disclosures: The author reported no conflicts of interest.

The *Journal* policy requires editors and reviewers to disclose conflicts of interest and to decline handling or reviewing manuscripts for which they may have a conflict of interest. The editors and reviewers of this article have no conflicts of interest.

Received for publication April 21, 2021; revisions received April 21, 2021; accepted for publication April 22, 2021; available ahead of print May 1, 2021.

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J Thorac Cardiovasc Surg 2022;163:209-10

0022-5223/\$36.00

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<https://doi.org/10.1016/j.jtcvs.2021.04.069>

Consider:

1. Symptomatic TOF in patients <2.5 kg is worse than most of us would have admitted. Other recent publications have also been sobering in this regard.^{2,3} This study, from leading pediatric heart programs, is concerning—at 5 years of follow-up, whether a child is treated with an initial palliation or a primary repair, their chance of being dead is 15% to 18%. This is not at all how we counsel families.
2. We are all over the map in treatment of TOF, and it is likely that this will get worse before it gets better. The studied cohort of catheter-based therapies for TOF palliation were largely just balloon pulmonary valve