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Commentary: Attrition after superior cavopulmonary connection; we don't have to be perfect but better than yesterday

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CENTRAL MESSAGE

In children with HLHS who fail to progress following SCPC, few opportunities exist to modify their course short of improving their transplant candidacy and support while on transplant waiting list.

In the current issue of the *Journal*, Lawrence and colleagues¹ from Philadelphia review their large cohort of 856 children born with hypoplastic left heart syndrome (HLHS) who survived to hospital discharge following superior cavopulmonary connection (SCPC). They report the progression of these children subsequent to SCPC and focus on those who failed to move toward undergoing the Fontan operation. They found that interstage attrition (between SCPC and Fontan) occurred in 71 children (8.3%) including 52 (6.1%) who died, 12 (1.4%) who received or were waiting for heart transplantation, and 7 (0.8%) who were alive but not deemed suitable candidates for Fontan. They examined risk factors associated with this attrition and found that the use of right ventricle to pulmonary artery (RVPA) shunt at Norwood, atrioventricular valve repair or performance of other procedures at SCPC, cardiopulmonary bypass duration at SCPC, and length of stay after SCPC to be significant risk factors. Importantly, they found that this attrition rate was not affected by era in this study that spanned over 30 years. They inferred that strategies to improve survival in this interstage period are needed.¹

Multistage palliation management of children with HLHS continues to evolve, and advances in all aspects in the care of these challenging patients have resulted in a remarkable improvement of their outlook.² Much of the emphasis has been on the greatest-risk period that includes

perioperative care and interstage period between Norwood and SCPC.² The findings from the authors' experienced, large center are significant and reflect that despite our success in decreasing early death, we are unable to sustain survival toward the final palliation stage in a number of children, and that we have failed to make a marked improvement in the outlook of these children during that stage over the past 3 decades.¹

In our continuous task to advance the management of children with complex congenital heart defects, we should constantly examine opportunities to improve survival and life quality. In the case of interstage attrition between SCPC and Fontan, the question is: are there any potentially correctable factors that would allow us to decrease this attrition and increase the total number of children who successfully complete their multistage palliation strategy and receive the Fontan operation? However, this interstage attrition between SCPC and Fontan might represent the reality of HLHS that is not necessarily modifiable, as there will always be a number of children with anatomic or hemodynamic abnormalities that limit their Fontan candidacy. In that case, the question becomes: what strategies can we apply to increase the proportion of marginal children who receive heart transplantation instead of dying subsequent to undergoing SCPC?

To address the first question, the authors suggest that the use of RVPA shunt at Norwood and performance of

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additional procedures at SCPC including atrioventricular valve repair are risk factors for attrition after SCPC. They further suggest that failure to improve this attrition in the recent era might be related to the relative increase use of RVPA shunt.¹ However, is that enough evidence against the use of RVPA shunt? Other series, including the Single Ventricle Reconstruction trial, similarly showed that despite the greater early survival with the use of RVPA shunt as opposed to the modified Blalock–Taussig shunt, the eventual survival balances within few years from Norwood with more attrition in the RVPA shunt group.³ This later death might be attributed to the effect of right ventriculotomy on right ventricular function, although that has not been clearly demonstrated in published reports, including the current one by Lawrence and colleagues. Given that many surgeons and centers have achieved improved early survival following Norwood with RVPA shunt use, it is unlikely that many will be convinced to take a step back toward the exclusive use of the Blalock–Taussig shunt again.^{3–5} Whether recent RVPA shunt modifications such as the use of the dunk technique with spreading rather than resection of right ventricular muscles proves to be beneficial remains to be seen.⁶

As for additional procedures at time of SCPC including atrioventricular valve repair, that is also a complicated issue. Pulmonary artery augmentation is commonly needed at SCPC, as pulmonary artery hypoplasia is seen infrequently due to stretching, compression, or distortion effects. Surgeons have tried various techniques at time of Norwood to promote pulmonary artery growth and have not identified so far a superior technique that obviated the need for pulmonary artery work.⁷ In addition, atrioventricular valve regurgitation is unfortunately also often seen and can be due to organic deformity rather than functional manifestation. Consequently, avoidance of valve repair or replacement might not always be possible.^{8,9} Therefore, to answer the first question, it seems that opportunities to decrease total attrition and increase the number of patients who reach Fontan stage successfully are limited, since there are no truly modifiable risk factors to improve on.

Consequently, it seems to me that the best chance to improve survival in these children with anatomic and hemodynamic substrates that limit their Fontan candidacy is to increase their odds to receive heart transplantation (being the one currently valid alternative to multistage palliation strategy). Current data suggest that these children are often not considered transplant candidates due to clinical, immunologic and occasionally anatomic factors. In addition, they suffer from relatively high waiting-list mortality, and some of them have lower post-transplant survival.^{10,11} There are

ample opportunities to improve the transplantation outlook of these patients, including early identification and listing of proper candidates, better medical and mechanical support of these children while awaiting proper donors, improved immunosuppression strategies and management of sensitized patients, and hybrid interventions that could temporarily improve condition of patients awaiting transplantation (for example, tricuspid valve replacement using a percutaneous valve that I used myself in handful of cases with various success).

In summary, the reality is that a number of children with HLHS will continue to be poor candidates for Fontan and there are not many modifiable factors to influence this number. However, to provide better service to these complex children, our focus should be on early identification and better management of these patients to receive the one valid alternative strategy that we currently have at our disposal and that is heart transplantation.

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