stage 2 operation before 3 months of age.⁹ Although the youngest patients in this series all survived, a Blalock-Taussig shunt or Sano conduit may have been a better option.

In the end, the study provides an interesting look at an unusual scenario uncommon to most centers. This novel experience shows that a combined Norwood CPS is a viable option in late-presenting infants with increased pulmonary blood flow. Although the postoperative course may be rocky in this challenging situation, often requiring delayed sternal closure and pulmonary vasodilator therapy, intermediate survival appears comparable to the conventional sequence of staged operations.^{1,2,4}

References

- Ohye RG, Sleeper LA, Mahony L, Newburger JW, Pearson GD, Lu M, et al. Comparison of shunt types in the Norwood procedure for single-ventricle lesions. *N Engl J Med*. 2010;362:1980-92.
- Meza JM, Hickey E, McCrindle B, Blackstone E, Anderson B, Overman D, et al. The optimal timing of stage-2-palliation after the Norwood operation. *Ann Thorac Surg*. 2018;105:193-9.
- Ismail MF, Elmahrouk AF, Arafat AA, Hamouda TE, Alshaikh BA, Shihata MS, et al. Evolution of the Norwood operation outcomes in patients with late presentation. *J Thorac Cardiovasc Surg*. 2020;159:1040-8.
- Elmahrouk AF, Ismail MF, Arafat AA, Dohain AM, Edrees AM, Jamjomm AA, et al. Combined Norwood and cavopulmonary shunt as the first palliation in late presenters with hypoplastic left heart syndrome and single-ventricle lesions. *J Thorac Cardiovasc Surg*. 2022;163:1592-600.
- Lai L, Laussen PC, Cua CL, Wessel DL, Costello JM, del Nido PJ, et al. Outcomes after bidirectional Glenn operation: Blalock-Taussig shunt versus right ventricle-to-pulmonary artery conduit. *Ann Thorac Surg*. 2007;83:1768-73.
- Adatia I, Atz AM, Wessel DL. Inhaled nitric oxide does not improve systemic oxygenation after bidirectional superior cavopulmonary anastomosis. *J Thorac Cardiovasc Surg*. 2005;129:217-9.
- Agarwal HS, Churchwell KB, Doyle TP, Christian KG, Drinkwater DC, Byrne DW, et al. Inhaled nitric oxide use in bidirectional Glenn anastomosis for elevated Glenn pressures. *Ann Thorac Surg*. 2006;81:1429-35.
- Cua CL, McConnell PI, Meza JM, Hill KD, Zhang S, Hersey D, et al. Hybrid palliation: outcomes after the comprehensive stage 2 procedure. *Ann Thorac Surg*. 2018;105:1455-60.
- Galantowicz M, Yates AR. Improved outcomes with the comprehensive stage 2 procedure after an initial hybrid stage 1. *J Thorac Cardiovasc Surg*. 2016;151:424-9.

See Article page 1592.

Check for updates

Commentary: The right procedure for the right patient

Maryam A. Al-Omair, MD, and Christoph Haller, MD

The Achilles' heel of single-ventricle palliation is the inherently unstable interstage circulation following both the Norwood procedure with systemic-to-pulmonary artery shunt or stage I hybrid palliation. This is reflected in the high mortality reported in patients with hypoplastic left heart syndrome before completion of a cavopulmonary shunt and the subsequent flattening of survival curves after stage II palliation.¹ Primary construction of

Maryam A. Al-Omair, MD, and Christoph Haller, MD

CENTRAL MESSAGE

Primary combined Norwood cavopulmonary shunt procedures are feasible, but only in a highly selected group of patients.

a cavopulmonary anastomosis is impeded by high pulmonary vascular resistance (PVR) in neonates and young infants. Attempts to achieve a primary in-series circulation showed that this strategy was challenging, although not impossible.²⁻⁴

Elmahrouk and colleagues⁵ report their experience with primary combined Norwood and cavopulmonary shunt procedures in late-presenting single-ventricle type patients. The case series includes all patients aged 6 weeks or older who underwent a Norwood procedure with cavopulmonary

From the Department of Cardiovascular Surgery, The Labatt Family Heart Centre, The Hospital for Sick Children, University of Toronto, Toronto, Ontario, Canada. 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 Nov 6, 2021; revisions received Nov 6, 2021; accepted for publication Nov 8, 2021; available ahead of print Nov 15, 2021.

Address for reprints: Christoph Haller, MD, Division of Cardiovascular Surgery, The Hospital for Sick Children, 555 University Ave, Toronto, Ontario M5G 1X8, Canada (E-mail: christoph.haller@sickkids.ca).

J Thorac Cardiovasc Surg 2022;163:1602-3

0022-5223/\$36.00

Copyright © 2021 by The American Association for Thoracic Surgery

<https://doi.org/10.1016/j.jtcvs.2021.11.018>

shunt irrespective of the underlying cardiac defect, with 50% of patients having a dominant right ventricle. They report a 75% survival rate with a maximum follow-up of 4 years. Deaths as a result of cardiac reasons appeared to be primarily driven by high PVR.

The setting of this major tertiary care center exposes the authors to a unique patient cohort of late-presenting single-ventricle patients. Late diagnosis and limited access to health care force the group to innovate and adopt palliative strategies that would otherwise rarely be necessary. The case series is an extension of previously reported results by the same group adding patients treated at the King Abdulaziz University Hospital.⁶ Clearly, there is an inherent selection process at work because patients had to maintain a certain degree of hemodynamic stability to survive weeks of uncontrolled in-parallel circulation. Ductal patency was present in all patients despite only 2 receiving preoperative prostaglandin infusions. Despite high Qp/Qs ratios, oxygen saturations were not excessive, ranging in the high 80s. PVR was elevated at baseline, but responsive to oxygen. Not unexpectedly, death was reported primarily in patients with elevated PVR: 1 patient with known preoperatively elevated PVR, the other needing cavopulmonary shunt take-down postoperatively.

The authors can be congratulated on an excellent series highlighting a very challenging group of patients and good postoperative outcome. As with so many other physiologic aspects of single-ventricle palliation, the remaining questions arise in those patients who are borderline—in this series at the younger end of the spectrum—presenting late for conventional stage I palliation but early enough that the pulmonary vascular bed may not yet be receptive for a cavopulmonary shunt. Looking closer at the authors' previous report on Norwood procedure outcomes, there is a small group of patients with overlapping strategies; that is, patients aged approximately 6 weeks who were treated with Norwood Blalock–Taussig/Sano shunt or Norwood cavopulmonary shunt.⁶ It would be most valuable to identify factors that favored 1 strategy over the other. The

report also omits those patients who presented late but were not considered candidates for a combined Norwood cavopulmonary shunt strategy. Were those patients offered a palliative strategy at all? Similarly, should we consider a primary Norwood cavopulmonary strategy in even younger patients if pulmonary blood flow and vascular resistance are reassuring?

Challenges push innovations and many surgical strategies applied today have been born from necessity. Elmahrouk and colleagues⁵ show encouraging results with a combined Norwood cavopulmonary shunt strategy in late presenting single-ventricle patients. However, a primary cavopulmonary shunt strategy has been successfully reported in patients aged ≥ 6 weeks by others as well.³ The true challenge lies in the management of patients to reach the necessary age and level of maturity of the pulmonary vascular bed to render them suitable for this strategy—a process with significant selection along the way.

References

1. Newburger JW, Sleeper LA, Gaynor JW, Hollenbeck-Pringle D, Frommelt PC, Li JS, et al. Transplant-free survival and interventions at 6 years in the SVR trial. *Circulation*. 2018;137:2246-53. <https://doi.org/10.1161/CIRCULATIONAHA.117.029375>
2. Norwood WI, Kirklin JK, Sanders SP. Hypoplastic left heart syndrome: experience with palliative surgery. *Am J Cardiol*. 1980;45:87-91. [https://doi.org/10.1016/0002-9149\(80\)90224-6](https://doi.org/10.1016/0002-9149(80)90224-6)
3. Reddy VM, Liddicoat JR, Hanley FL. Primary bidirectional superior cavopulmonary shunt in infants between 1 and 4 months of age. *Ann Thorac Surg*. 1995;59:1120-6. [https://doi.org/10.1016/0003-4975\(95\)00014-C](https://doi.org/10.1016/0003-4975(95)00014-C)
4. Slavik Z, Lamb RK, Webber SA, Webber SA, Devlin AM, Keeton BR, et al. Bidirectional superior cavopulmonary anastomosis: how young is too young? *Heart*. 1996;75:78-82. <https://doi.org/10.1136/hrt.75.1.78>
5. Elmahrouk AF, Ismail MF, Arafat AA, Dohain AM, Edrees AM, Jamjomm AA, et al. Combined Norwood and cavopulmonary shunt as the first palliation in late presenters with hypoplastic left heart syndrome and single-ventricle lesions. *J Thorac Cardiovasc Surg*. 2022;163:1592-600.
6. Ismail MF, Elmahrouk AF, Arafat AA, Hamouda TE, Alshaikh BA, Shihata MS, et al. Evolution of the Norwood operation outcomes in patients with late presentation. *J Thorac Cardiovasc Surg*. 2020;159:1040-8. <https://doi.org/10.1016/j.jtcvs.2019.07.154>