Commentary: It takes a village: Changing the trajectory of pulmonary vein stenosis outcomes

Rachel D. Vanderlaan, MD, PhD,^a and Osami Honjo, MD, PhD^{a,b}

From the ^aDivision of Cardiovascular Surgery, The Hospital for Sick Children, Toronto; and ^bDepartment of Surgery, University of Toronto, Toronto, Ontario, Canada.

Disclosures: Authors have nothing to disclose with regard to commercial support.

Received for publication Sept 19, 2019; revisions received Sept 19, 2019; accepted for publication Sept 20, 2019; available ahead of print Nov 9, 2019.

Address for reprints: Osami Honjo, MD, PhD, Division of Cardiovascular Surgery, The Hospital for Sick Children, 555 University Ave, Toronto, Ontario, M5G 1X8, Canada (E-mail: osami.honjo@sickkids.ca).

J Thorac Cardiovasc Surg 2020;159:1037-8

0022-5223/\$36.00

Copyright © 2019 by The American Association for Thoracic Surgery

<https://doi.org/10.1016/j.jtcvs.2019.09.100>

Accelerating improvements in clinical care for children with PVS.

Central Message

Improvement in outcomes in pulmonary vein stenosis (PVS) can be accelerated through institutional team-based approaches and with multi-institution collaboration through PVS Network initiatives.

See Article page 1029.

Primary pulmonary vein stenosis (PVS) remains a formidable challenge to clinicians, as mortality and rates of re-intervention remain high. In the accompanying article, Rosenblum and colleagues¹ present their contemporary institutional outcomes for children with primary PVS. Using the PVS score applied by Kalfa and colleagues, the data from the Emory group confirm previous reports regarding poor outcomes for patients with bilateral PVS compared with patients with unilateral disease.²⁻⁴ Their results also highlights that the majority (91%) of children in their study had 2 or fewer pulmonary veins and were typically unilateral PVS (as opposed to bilateral).

These results are of interest with respect to practice patterns, as what is unknown is the denominator or spectrum of PVS disease burden at their institution—a limitation of most surgical retrospective studies. Surgical bias can account for a greater likelihood to operate on children with less severe PVS,⁴ whereas children with progressive multi-vessel disease (>2, bilateral) or other high-risk comorbidities can more likely be directed toward a palliation pathway, given the historical poor prognosis. In contrast, a low institutional burden of advanced PVS disease could represent an aggressive anticipatory surveillance strategy whereby disease is caught early and interventions done expeditiously. Although not the focus of their surgical outcomes manuscript, the former challenges us to understand the denominator of PVS at our own institutions, and the latter challenges us to review how we approach care for children with PVS.

Although the Emory results may not be a resounding improvement from previously published results for children with multivessel disease,^{5,6} the results highlight the need to refocus our attention to children with PVS. Advancement in PVS clinical outcomes seems at times stagnant and unlikely to improve in the absence of new technology or therapy; however, in this current landscape, advancements in clinical care for children with PVS can be accelerated; care can be

optimized at the institutional level and across institutions with the development of the multi-institutional collaborations.

OPTIMIZING CARE AT THE INSTITUTIONAL LEVEL

At the institutional level, there is a need to consolidate expertise to accelerate learning in this relatively rare disease. The clinical heterogeneity of disease presentation and varied treatment practices across primary caregivers can make advancements in institutional outcomes difficult to discern. As a result, institutions such as Toronto, Boston, Texas, and Emory have implemented a multidisciplinary team dedicated to following children with PVS. These teams consist of surgeons, interventionalists, cardiologists, and radiologists who review cases and help guide management of patients, thereby consolidating and harmonizing institutional experience.

The use of clinical surveillance protocols by institutions is another way to accelerate understanding of disease progression, given the frequently clinically silent progression of PVS. By instituting structured follow-up, teams are better equipped to understand the velocity of disease progression and likewise intervene on early disease, before disease becomes diffuse in upstream vessels and is no longer amenable to intervention.³ Ideally, treatment plans should be anticipatory, not reactionary.

The PVS team-based approach highlights that a single intervention will not provide a definitive cure in most children with multivessel disease. Recurrence rates are high,

and vigilance with clinical surveillance tools such as echocardiography, computed tomography/magnetic resonance imaging, and diagnostic catheterization to detect early disease progression are optimized in a team-based approach, where the appropriate surveillance modality can be applied for the given clinical scenario. Likewise, the use of surgery, stents, balloon angiography, and experimental therapies may all be required at different points within a child's life to reduce the burden of disease, and thereby necessitate collaboration between cardiologists, surgeons, and interventionalists. In our institution, inclusion of lung transplant team in PVS rounds has helped to identify children with poor prognosis and help design bridging strategies to lung transplant.⁷

MOVING TOWARD CONSENSUS

The push to advance outcomes in PVS through multi-institutional collaboration has become a central focus with the inception of the PVS Network (www.pvsnetwork.org), a collective of institutions dedicated to improving research and clinical care for children with PVS. The PVS Network has compiled the first North American registry, with more than 500 children with a diagnosis of PVS registered, and will provide insights into surveillance and treatment practices across institutions and help to facilitate consensus to different aspects of management for children with PVS. With the first full-day dedicated PVS Network Symposium occurring in conjunction with the Neonatal and Childhood Pulmonary Vascular Disease meeting in the spring of 2020, cardiologists, surgeons, interventionalists, pulmonary hypertension specialists, and researchers will have the opportunity to identify key priorities of the PVS community for the coming years. Through creation of a strong

multi-institutional infrastructure and a historical contemporary control cohort, the PVS community is poised to implement prospective studies that will attempt to address current knowledge gaps and improving our understanding of PVS in distinct populations.

Incremental progress in rare diseases is difficult to ascertain at an institutional level and necessitates collaboration across disciplines and institutions. Rosenblum and colleagues are commended for their multidisciplinary approach to management and follow-up surveillance of children with PVS. We are challenged to understand the spectrum of PVS both at an institutional and multi-institutional level to better understand how to optimize clinical care. Through collaboration, we can change the trajectory of children with PVS and improve clinical outcomes.

References

1. Rosenblum JM, Altin HF, Gillespie SE, Bauser-Heaton H, Kanter KA, Sinha R, et al. Management outcomes of primary pulmonary vein stenosis. *J Thorac Cardiovasc Surg.* 2020;159:1029-36.e1.
2. Kalfa D, Belli E, Bacha E, Lambert V, di Carlo D, Kostolny M, et al. Primary pulmonary vein stenosis: outcomes, risk factors, and severity score in a multicentric study. *Ann Thorac Surg.* 2017;104:182-9.
3. Seale AN, Webber SA, Uemura H, Partridge J, Roughton M, Ho SY, et al. Pulmonary vein stenosis: the UK, Ireland and Sweden collaborative study. *Heart.* 2009;95:1944-9.
4. Lo Rito M, Gazzaz T, Wilder TJ, Vanderlaan RD, Van Arsdell GS, Honjo O, et al. Pulmonary vein stenosis: severity and location predict survival after surgical repair. *J Thorac Cardiovasc Surg.* 2016;151:657-66.e2.
5. Quinonez LG, Gauvreau K, Borisuk M, Ireland C, Marshall AM, Mayer JE, et al. Outcomes of surgery for young children with multivessel pulmonary vein stenosis. *J Thorac Cardiovasc Surg.* 2015;150:911-7.
6. Viola N, Alghamdi AA, Perrin DG, Wilson GJ, Coles JG, Caldarone CA. Primary pulmonary vein stenosis: the impact of sutureless repair on survival. *J Thorac Cardiovasc Surg.* 2011;142:344-50.
7. Galasso M, Vanderlaan RD, Solomon M, Caldarone C, Keshavjee S, Cypel M, et al. A successful lung transplant from a 3-year-old donor after controlled cardiac death followed by ex vivo lung perfusion: a case report. *J Thorac Cardiovasc Surg.* 2019;158:e149-52.