



Published in final edited form as:

J Card Surg. 2022 September ; 37(9): 2651–2652. doi:10.1111/jocs.16651.

Commentary: Should We “Keep Rollin’” for PA/VSD/MAPCA?

Raymond J. Strobel, MD, MSc¹, Andrew M. Young, MD¹, Irving L. Kron, MD¹

¹Division of Cardiac Surgery, Department of Surgery, University of Virginia, Charlottesville, VA

Abstract

Pulmonary atresia with ventricular septal defect and major aortopulmonary collateral arteries (PA/VSD/MAPCA) represents an anatomically diverse and technically demanding spectrum of congenital disease. Here, we review a manuscript by Onalan and colleagues in the *Journal of Cardiac Surgery* detailing a retrospective, single-center cohort study of patients undergoing unifocalization for PA/VSD/MAPCA via either a pulmonary artery patch augmentation or pericardial roll technique. While they report statistically equivalent outcomes using both techniques, longer follow-up and increased sample size are necessary to determine efficacy and safety.

Commentary:

In this month’s *Journal*, Onalan and co-authors¹ report on a retrospective, single-center cohort study of patients undergoing unifocalization of pulmonary atresia, ventricular septal defect, and major aortopulmonary collateral arteries. A total of 48 patients were included from 2015 to 2021, 14 (29.2%) of which underwent single-stage complete unifocalization, 26 (54.1%) unifocalization with concomitant systemic to pulmonary artery shunt placement with subsequent consideration for complete repair, and 8 underwent initial systemic-pulmonary or aortopulmonary window creation without concomitant unifocalization. Regarding technique, 75% (30) of patients received a unifocalization featuring a classic side-to-side anastomosis of MAPCAs to the native pulmonary artery with pericardial patch augmentation. The remaining 25% (10) of patients received a circumferential pericardial roll to which both the MAPCAs and unifocalized right and left native PAs were anastomosed. Ultimately, most patients achieved a complete repair (32, 66.6%). Overall mortality was 16.6% (8 deaths), and outcomes (i.e., mortality, reintervention rate, length of stay, perioperative RV/LV pressure ratio) between pericardial roll and pericardial patch augmentation groups were not statistically different. The length of follow-up was 0.9 years (0.1–3) in the pericardial roll group and 2.6 years (0.1–6) in the patch augmentation group.

We (Figure 1) commend the authors for their excellent outcomes in this challenging patient population. While comparisons between the pericardial roll and pericardial patch

Corresponding Author: Irving L. Kron, MD, Division of Thoracic and Cardiovascular Surgery, Department of Surgery, 1215 Lee St., PO Box 800679, Charlottesville, VA 22908, ILK@virginia.edu.

Conflict of Interest: none

Disclosures: The authors have nothing to disclose

IRB approval and informed consent: N/A

augmentation groups were statistically non-significant, median follow-up is relatively short (0.9 years and 2.6 years, respectively). This may be of material importance for outcomes like median freedom from PA reintervention, which appeared to trend towards significance favoring the patch augmentation technique (6 months vs. 13 months, $p = 0.077$), especially given concerns that may be raised by the use of circumferential, non-viable conduit which is unable to grow along with the child. Longitudinal studies in this population will be critical to assessing this relatively new technique. Similarly, while the authors state that they prefer the roll technique for patients who "...had absent or small central pulmonary arteries," it is not entirely clear what size criteria are used to establish what is 'small enough' or how they manage the anastomosis between their RV-PA conduit and the roll itself. The authors' results suggest that the pericardial roll technique is worthy of continued study.

Advances in the surgical treatment of PA/VSD/MAPCA have allowed for excellent outcomes for those who can achieve a complete repair.² Using an algorithmic approach favoring single-stage unifocalization for this heterogeneous and anatomically complex patent population, Malhotra and Hanley reported an actuarial survival of 85.5% at five years follow-up. This must be compared to the natural history of this disease (50% survival at two years) and equivalent survival rates but significantly lower rates of complete repair in series detailing staged repair.³ The Stanford Group has described the "ideal outcome" for these patients as consisting of three key, measurable endpoints: (1) Survival, (2) Complete Repair, and (3) The RV/Ao pressure ratio is below a designated level postoperatively (i.e., 0.45).⁴ While comparatively less is known regarding the efficacy and safety of the pericardial roll technique described by Onalan and colleagues, if it empowers the congenital surgeon to achieve this "ideal outcome" while avoiding a higher reintervention rate, then the greater community will welcome it. As the authors rightly propose in their discussion, additional studies which feature large sample sizes and longer follow-up are needed to fully assess the pericardial roll technique for this complex patient population.

Funding Statement:

This work was supported by a research grant from NHLBI/NIH (T32HL007849). The content is solely the responsibility of the authors and does not represent the official views of the National Institutes of Health.

Data Availability Statement:

This invited commentary does not include any original data.

References:

1. Onalan MA, Cicek M, Rum M, Yurdakok O, et al. Unifocalization with Pericardial Roll Technique in Pulmonary Atresia with Ventricular Septal Defect and Major Aortopulmonary Collateral Arteries. *J Card Surg.* 2022. [In Press].
2. Malhotra SP, Hanley FL. Surgical management of pulmonary atresia with ventricular septal defect and major aortopulmonary collaterals: a protocol-based approach. *Semin Thorac Cardiovasc Surg Pediatr Card Surg Annu* 2009;145–51. 10.1053/j.pcsu.2009.01.017. [PubMed: 19349030]
3. Gupta A, Odum J, Levi D, Chang R-K, Laks H. Staged repair of pulmonary atresia with ventricular septal defect and major aortopulmonary collateral arteries: experience with 104 patients. *J Thorac Cardiovasc Surg* 2003;126:1746–52. 10.1016/s0022-5223(03)01200-5. [PubMed: 14688682]

4. Mainwaring RD, Patrick WL, Hanley FL. Surgical Management of Pulmonary Atresia With Ventricular Septal Defect and Major Aortopulmonary Collateral Arteries: Part II—Midline Unifocalization and Unifocalization Revision. *Operative Techniques in Thoracic and Cardiovascular Surgery* 2019;24:56–79. 10.1053/j.optechstcvs.2019.07.002.

Author Manuscript

Author Manuscript

Author Manuscript

Author Manuscript

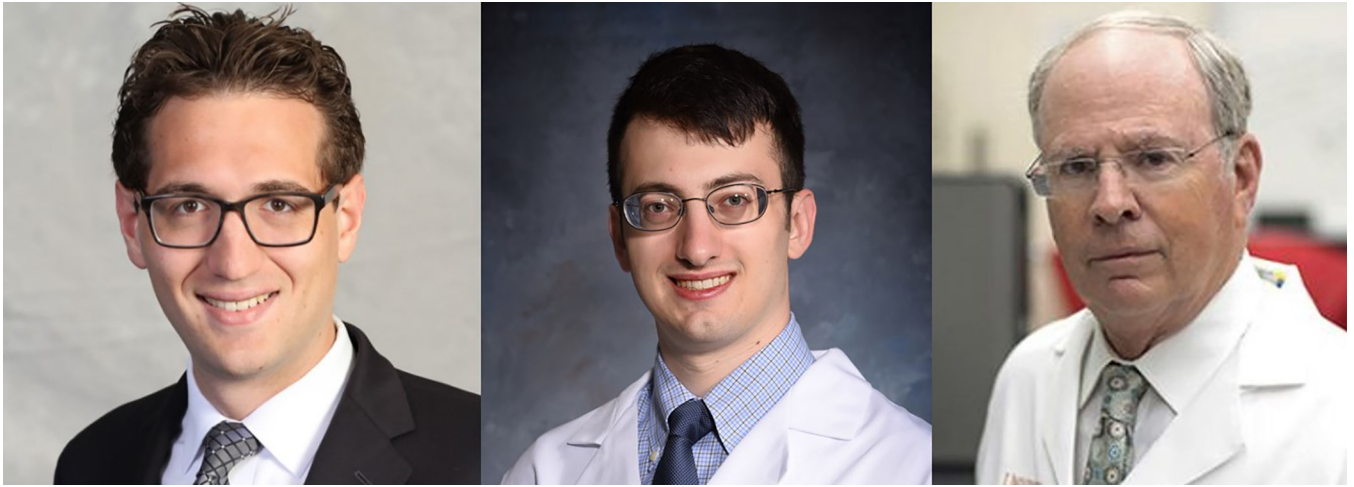


Figure:
Raymond J. Strobel, MD, MSc (left), Andrew M. Young, MD (center) and Irving L. Kron,
MD (right)