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Commentary: Management outcomes of primary pulmonary vein stenosis: What we know now and what we can expect in the future

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Primary pulmonary vein stenosis (PPVS) may be one of the most challenging cases in the pediatric cardiovascular diseases. Because of its rarity, PPVS is also one of diseases with which most pediatric cardiac cardiologists and surgeons lack experience. Even with its small population, the study by Rosenblum and colleagues¹ reported in this issue of the *Journal* represents the largest PPVS series reported to date.

The authors report unsatisfactory results with both primary surgical repair and initial percutaneous intervention. Nonetheless, the study contributes to our understanding of the treatment and management of PPVS. For example, nearly 82% patients underwent a sutureless technique with no improvement in outcomes, possibly demonstrating the need for modification of existing surgical techniques for PPVS and showing that novel surgical techniques or adjuvant medical therapy merit investigation. Moreover, the percutaneous outcomes of this study in high-risk patients are very disappointing, which indicates that primary surgical repair may be the preferred choice for PPVS. Finally, the authors demonstrate the effectiveness of PPVS severity score in identifying the risks of mortality and reintervention, highlighting the importance of multi-institutional studies in the future for developing a universal PPVS severity score.

One limitation that the authors do not mention is the positional relationship of the pulmonary veins with the atria,

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Primary pulmonary vein stenosis management still has a long way to go.

especially considering the >90% of patients in this cohort had an associated congenital heart disease. In our surgical experience, as well as that of other groups, the positional relationship between the pulmonary veins and bilateral atria is variable in total anomalous pulmonary venous connection (TAPVC), and this relationship is important for determining the optimal surgical approach.^{2,3} Some work is being done on using 3D printing technology to investigate the 3-dimensional positional relationship of pulmonary veins and the atria in complicated TAPVC cases before surgery. Because pulmonary veins are very susceptible to rotation and distortion of the anastomosis angle, it is reasonable to suppose that the 3-dimensional positional relationship of the pulmonary veins with the atria in PPVS may be one of main factors in the poor results, especially in cases with diffuse and long-term pulmonary venous stenosis. Thus, more information on the 3-dimensional positional relationship of pulmonary veins and the atria in this cohort of patients, as well as other multi-institutional PPVS patients, may help improve our understanding of this complex disease and determine the optimal surgical approach.

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