Pulmonary vein stenosis: Challenges ahead

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In this issue of the *Journal*, Quinonez and colleagues¹ report their experience with and treatment of 49 patients with pulmonary vein stenosis. This article takes on the very difficult challenge to map out the “relentless” nature of multivessel pulmonary vein stenosis and the multidisciplinary commitments that are required for its treatment. We are encouraged by their acceptable short-term survival—67% at 1 year—that is the result of good surgery and high awareness upon postoperative follow-up, where the threshold to catheterization and intervention was low. Low weight at surgery, early surgical repair, and somewhat intriguing right ventricle pressure <75% of systemic are considered risk factors with poor prognosis.

When multiple strategies employed to solve a single problem have persistently disappointing results and a heavy burden of repeated surgeries and interventions, we need to ask ourselves what is wrong.

Pulmonary vein stenosis is at its fundament a process of pathologic cell proliferation and deposition of large amounts of extracellular matrix.² Despite the rare occurrence and difficulties in retrieving human tissue for basic research, enlightening knowledge has been generated. Riedlinger and colleagues³ showed that platelet-derived growth factors A and B, vascular endothelial growth factor, fibroblast growth factor, and activated receptor tyrosine kinases are involved; and Kato and colleagues^{4,5} demonstrated that transforming growth factor beta-1, a stimulator of matrix expansion, is upregulated in human pulmonary vein stenosis. Is this type of basic science important? Are its findings important? What does it mean in terms of the management of recurrent pulmonary vein stenosis?

Accepting the shortcomings of surgery and catheter interventions, it was particularly uplifting to read the Discussion offered by Quinonez and colleagues, where it was revealed that 59% of the patients in their study were also being

enrolled in another trial to test the effect of imatinib and bevacizumab. The choices of these drugs were not made at random. The former inhibits tyrosine kinases, and the latter is an antibody that inhibits vascular endothelial growth factor. A brief comment on preliminary data reveals that the treatments had no effect on survival. Obviously, we still have many challenges ahead.

In the meantime, we can learn from Quinonez and colleagues’ purposeful and tenacious approach to a very difficult problem. These are the hallmarks of pediatric cardiac surgery. We shall wait with patience for the results of the coming studies.

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Central Message

Strategies to improve survival and good quality of life for patients with pulmonary vein stenosis face difficult challenges.

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