References

1. Lacour-Gayet F, Rey C, Planche C. Pulmonary vein stenosis. Description of a sutureless surgical procedure using the pericardium in situ [article in French]. *Arch Mal Coeur Vaiss*. 1996;89:633-6.
2. Najm HK, Caldarone CA, Smallhorn J, Coles JG. A sutureless technique for the relief of pulmonary vein stenosis with the use of in situ pericardium. *J Thorac Cardiovasc Surg*. 1998;115:468-70.
3. Yun TJ, Coles JG, Konstantinov IE, Al-Radi OO, Wald RM, Guerra V, et al. Conventional and sutureless techniques for management of the pulmonary veins: evolution of indications from postrepair pulmonary vein stenosis to primary pulmonary vein anomalies. *J Thorac Cardiovasc Surg*. 2005;129:167-74.
4. Shi G, Zhu Z, Chen J, Ou Y, Hong H, Nie Z, et al. Total anomalous pulmonary venous connection: the current management strategies in a pediatric cohort of 768 patients. *Circulation*. 2017;135:48-58.
5. Kalfa DK, Belli E, Bacha E, Lambert V, di Carlo D, Kostolny M, et al. Outcomes and prognostic factors for postsurgical pulmonary vein stenosis in the current era. *J Thorac Cardiovasc Surg*. 2018;156:276-86.
6. Vanderlaan RD, Rome J, Hirsch R, Ivy D, Caldarone CA. Pulmonary vein stenosis: treatment and challenges. *J Thorac Cardiovasc Surg*. 2021;161:2169-76.

See Article on page 2169.

Commentary: This commentary was challenging and relentless

Charles B. Huddleston, MD

Most reports on the subject pulmonary vein stenosis (PVS) begin with the admonition that this disease is challenging or relentless and Vanderlaan and colleagues¹ remind us once again of this simple fact. According to the Society of Thoracic Surgeons Congenital Heart Surgery database, there are approximately 150 operations performed annually in the United States and Canada for PVS in 115 centers. That means, of course, that most centers are performing 1 or 2 such operations annually. This underscores the importance of developing a network of interested parties to study this diagnosis. Vanderlaan and colleagues¹ represent such a group and provide a valuable addition to the literature and summarize the knowledge to date. What exactly can we learn by reading their article?

- Do we know what causes PVS? The answer is no. Apart from some interesting theoretical genetic, embryologic, and mechanical propositions the etiology of PVS remains elusive.

From the St Louis University School of Medicine, St Louis, Mo.

Disclosures: The author 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 June 13, 2020; accepted for publication June 14, 2020; available ahead of print June 29, 2020.

Address for reprints: Charles B. Huddleston, MD, St Louis School of Medicine, 1465 S Grand Blvd, St Louis, MO 63104 (E-mail: Charles.huddleston@health.slu.edu). *J Thorac Cardiovasc Surg* 2021;161:2178-9

0022-5223/\$36.00

Copyright © 2020 by The American Association for Thoracic Surgery

<https://doi.org/10.1016/j.jtcvs.2020.06.067>

Charles B. Huddleston, MD

CENTRAL MESSAGE

Pulmonary vein stenosis is a challenging and relentless disease.

- Do we know how to make the diagnosis? The answer is yes! The recommendation is to start with an echocardiogram (no surprise there) and then move on to cardiac catheterization, computed tomography or magnetic resonance angiography and possibly nuclear medicine perfusion scans. That essentially runs the gamut of diagnostic tests available to heart specialists.
- Do we know how best to treat PVS? Sort of. Balloon dilatation and stenting of pulmonary veins were once considered hopeless and harmful.² However, stents have made a comeback and drug-eluting stents appear better than bare-metal stents, recognizing that the largest drug-eluting stent is 5 mm. What is the best operation? The so-called sutureless technique (it actually does require sutures) has better midterm results than other techniques such as patch venoplasty, ostial resection, reimplantation, or unroofing. However, the recurrence rate and mortality remain somewhat high.

In addition, upstream stenosis proximal to the pericardial reflection would require a much more complex operation or intraoperative stent placement. After all, PVS is challenging and relentless.

- What are the indications for stent placement or surgery? That simple question is rarely answered in articles regarding PVS and unfortunately Vanderlaan and colleagues¹ do not provide any insight. A symptomatic patient with severe stenosis involving all 4 pulmonary veins and pulmonary hypertension clearly should have an intervention. But what about asymptomatic patients with normal right ventricular pressure and a single stenotic pulmonary vein?
- What about medical therapy? A few clinical trials of drugs have been performed. Those drugs that may hold promise are imatinib, bevacizumab, and losartan.

However, the studies are hampered by small numbers of patients and lack of controls.

- What if nothing is working? Lung transplantation remains the final solution, recognizing that the 5-year survival is around 50%.

What do we conclude from this? Perhaps a new, innovative procedure will be developed. Perhaps 1 of these drugs or something else will be found to be a difference maker. At the end of the day, PVS is challenging and relentless.

References

1. Vanderlaan RD, Rome J, Hirsch R, Ivy D, Caldarone CA. Pulmonary vein stenosis: treatment and challenges. *J Thorac Cardiovasc Surg*. 2021;161:2169-76.
2. Peng LF, Lock JE, Nugent AW, Jenkins KJ, McElhinney DB. Comparison of conventional and cutting balloon angioplasty for congenital and postoperative pulmonary vein stenosis in infants and young children. *Cath Cardiovasc Int*. 2010;75:1084-90.