Commentary: This commentary was challenging and relentless

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Most reports on the subject pulmonary vein stenosis (PVS) begin with the admonition that this disease is challenging or relentless and Vanderlaan and colleagues 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 colleagues 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.

- 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. 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.

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In addition, upstream stenosis proximal to the pericar- 
dial 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 col-
leagues do not provide any insight. A symptomatic pa-
tient 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 ste-
notic 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 re-
mains the final solution, recognizing that the 5-year sur-
vival is around 50%.

What do we conclude from this? Perhaps a new, innova-
tive 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.

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