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Commentary: Relentless disease, relentless measures: Treat recurrent pulmonary vein obstruction early and often

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The constellation of total anomalous pulmonary venous connection (TAPVC), heterotaxy, a single ventricle, and primary pulmonary venous obstruction has long been recognized as an unfortunate set of cards.¹ Although some series have shown improving surgical outcomes over time, others have not.^{2,3} Nevertheless, despite the challenges of this difficult and sometimes recalcitrant form of congenital heart disease, the battle wages on in pursuit of better patient survival.

Spigel and colleagues⁴ present a large, single-institution series of patients with TAPVC, focusing on the influence of heterotaxy and preoperatively obstructed venous connection on subsequent mortality and recurrent obstruction. Compared with those without heterotaxy, TAPVC patients with heterotaxy demonstrated significantly worse survival, although much of this disparity may be attributed to the high incidence (90%) of single-ventricle anatomy within the heterotaxy group. Not surprisingly, overall survival was higher for TAPVC patients with 2 ventricles rather than single-ventricle anatomy (94% vs 63%).

Recurrent pulmonary vein obstruction represented another risk factor for mortality. As reported previously,^{5,6} recurrent vein disease predominantly presented within the first year after surgery. Similar to another study that identified heterotaxy, a single ventricle, and primary obstruction as risk factors for recurrent obstruction,⁶ this report showed primary

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Although recurrent pulmonary vein obstruction after TAPVC repair presents a difficult challenge, aggressive surveillance and intervention can lead to favorable outcomes.

obstruction with either heterotaxy or a single ventricle to be associated with increased pulmonary vein interventions. Notably, the need for pulmonary vein intervention was a marker for decreased survival. According to institutional practice, patients with heterotaxy and a single ventricle underwent more frequent surveillance, a bias that may have contributed to more interventions for these patients. However, this concern is mitigated by the observation that both heterotaxy and single-ventricle patients without primary obstruction did not experience increased interventions.

Perhaps the most interesting aspect of this study involves the outcomes of intervention for recurrent pulmonary vein obstruction. If there is a potential silver lining, it may be that disease recurrence appears to be fairly quiescent beyond the first year, resulting in a flattened curve that yielded a steady state for most patients.⁴ If detected and treated early and often, the fire of pulmonary vein disease can, in many patients, be eventually extinguished. Unfortunately, this success is not universal. For patients experiencing recurrent disease, each successive round resulted in increasingly worse (shorter) reintervention-free survival, suggesting the pathologic obstructive process was accelerating rather than slowing.

Accordingly, Spigel and colleagues⁴ recommend intense surveillance and intervention. With such a policy, they suggest that most patients can be rescued from the cycle of recurrent vein obstruction. Previously, in light of the relentless nature of the obstructive process and associated dismal

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prognosis,⁷ some may have considered such aggressive care with multiple reoperations to be futile or unwarranted, a sentiment that may have hindered the opportunity for repeat surgery in some cases. This study appears to argue instead for an escalation of care and for a greater willingness to perform serial operations; it advocates for staying in the fight. Hippocrates long ago counseled that extreme disease calls for extreme measures.⁸ In this case, relentless disease may call for relentless measures.

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Commentary: Total anomalous pulmonary venous connection, heterotaxy and pulmonary vein obstruction: The relentless pursuit of imperfection

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Spigel and colleagues¹ describe the medium-term outcomes among patients with total anomalous pulmonary venous connection (TAPVC) over nearly 25 years at their

Anusha Jegatheeswaran, MD, PhD, FRCSC (left), and Tara Karamlou, MD, MSc (right)

CENTRAL MESSAGE

Complex statistics can provide increased understanding of CHD patients but must be coupled with optimum research methodology to ensure we are not in relentless pursuit of an imperfect understanding.

institution. Their objective was to examine time-related mortality and reintervention in those with and without heterotaxy, with each group stratified by the presence or absence of pulmonary vein obstruction (PVO). The population consisted of 118 patients with heterotaxy (of whom 49 had PVO) and 218 patients without heterotaxy (of whom 87 had PVO). Not surprisingly, the authors found that patients with heterotaxy had greater mortality than those without and that those with PVO had worse survival than those

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