Pulmonary venous obstruction in the single ventricle patient: Possible causes, possible solutions

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See related article on pages 1332-8.

A recurring theme in the treatment of congenital heart disease is that surgical rearrangement of native anatomy provides early benefit but may also lead to future unanticipated circulatory problems that compromise the outcome of the initial surgery. Careful surveillance of patients who have undergone operation is critical to the refinement of surgical strategies and treatment of subsequent problems. This is one area in which congenital heart specialists do rather well. One example is the recognition that atrial enlargement after atriopulmonary Fontan-type connections can cause late right pulmonary vein compression, limiting Fontan repair longevity. Recognition of this complication was one factor that prompted the now universal shift toward more durable total cavopulmonary-type Fontan modifications. In this month's *Journal*, Kotani and colleagues¹ from the Hospital for Sick Children highlight an important but underrecognized form of unilateral pulmonary venous obstruction (PVO) that occurs in a small fraction of patients with single-ventricle physiology. The Toronto group has made a large number of contributions to the field of congenital heart surgery, most of which have come from their attention to meticulous long-term patient follow-up. Of particular note, this group has made innovative advances in the management of various forms of PVO,² and their current work represents another significant contribution to this area.

Kotani and colleagues¹ evaluated patients with single-ventricle anatomy born with normal pulmonary venous connections in which PVO developed at various times after initial surgical intervention as they progressed along the palliative pathway (almost all before Fontan procedure). Of 494 consecutive patients who underwent surgical interventions initiated with the intent to achieve eventual Fontan circulations, 24 (4.9%) had development of left-sided PVO, and involvement of the lower vein was

identified in 87%. Kotani and colleagues¹ limited their study to patients with left-sided PVO and hypothesized that compression from thoracic structures external to the veins is a primary etiology of left-sided PVO. They confined the study group to 15 patients with PVO for whom cardiac magnetic resonance (CMR) studies were available and compared them with 12 case-matched control subjects without PVO (matched for age, ventricular morphology, type of initial palliation, and stage of palliation) for whom CMR data were also available. Kotani and colleagues¹ appropriately note the limitations of the study. There is a paucity of literature on this patient group, and the current study provides the only comprehensive look at causative factors and patient outcomes to date. Findings highlighted by Kotani and colleagues¹ include the following: (1) Patients who have PVO develop are a high-risk group for single-ventricle palliation, with a roughly 50:50 chance of achieving successful Fontan repair. (2) Patients with PVO had increased cardiac area (relative to total thoracic area) and anterolateral positioning of the descending thoracic aorta relative to control subjects. (3) Echocardiography has poor sensitivity for the diagnosis of PVO. (4) Finally, "sutureless" PVO repair eliminated PVO in 4 of 7 patients in whom this was attempted. These findings implicate extrinsic vein compression in the pathogenesis of left-sided PVO. They also suggest that CMR should be used more widely as a surveillance and diagnostic tool to detect PVO or its potential substrate in patients with single-ventricle anatomy. Less costly available tools are inadequate for these purposes. Kotani and colleagues¹ do not overstate their case for the use of CMR in this patient group. It is to be hoped that follow-up work from this group and others will further clarify the appropriate role for CMR in these patients.

In addition to the highlighted findings, several addressed and unaddressed items merit attention. First, the presence of unilateral PVO alone does not independently portend failure of single-ventricle palliation. Thus the proverbial glass seems only half empty, considering the entire group of patients with single-ventricle anatomy and PVO. Many patients have adequate compensatory flow to the right lung develop, with pulmonary resistances that remain suitably low for Fontan palliation. Second, Kotani and colleagues¹ report that sutureless repairs were effective in relieving PVO in 4 of 7 patients. Unfortunately, they offer no data demonstrating therapeutic benefit to repair even when successful, and the role of PVO repair in this subset of patients is not demonstrated. With the low number of

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patients who have undergone repair, this weakness is understandable, and follow-up assessment of the effect and role for PVO repair in these patients would be of significant interest. Third, Kotani and colleagues¹ confined their assessment to patients with left-sided PVO. It would be useful to know whether any patients in their global cohort of patients with single-ventricle anatomy had right-sided PVO. In my limited experience, PVO in patients with single-ventricle anatomy born without anomalous connections has occurred solely on the left side. If PVO occurs almost exclusively on the left, the compressive pathogenesis postulated by Kotani and colleagues¹ and suggested by their data seems much more probable than the intrinsic venous disease that is postulated for other types of PVO. Understanding the pathogenesis of PVO is important, because a compressive etiology is more likely to be amenable to prevention or successful surgical relief if detected early in the course of disease.

As with right-sided PVO occurring after atriopulmonary Fontan repair, it is possible that at least some patients with left-sided PVO may have this phenomenon develop as a direct result of previous surgical intervention. Of the factors that Kotani and colleagues¹ correlated with the development of left-sided PVO, it is easily appreciated that previous palliation may induce volume overload and atrioventricular valve regurgitation, which result in cardiomegaly. The data of Kotani and colleagues¹ associate heart enlargement within the relatively limited left chest space with left-sided PVO. If correct, this provides yet another reason to limit the duration of volume overload and prevent or repair atrioventricular valve regurgitation early in the course of the disease. Less obvious is the possibility that the other identified factor, anterolateral position of the descending aorta, could also result from previous surgical palliation in some patients. Most congenital heart surgeons are aware that aortic arch reconstruction can be complicated by left bronchial compression. This complication is due to repositioning of the descending aorta. It is reasonable to question whether pulmonary venous compression could also occur as a result of aortic arch reconstruction in patients requiring this as a component of palliation. The Toronto group was among the first to advocate aggressive coarctation resection at stage 1 palliation and described an arch reconstruction that requires mobilization and advancement of the descending

aorta.² This technique has clearly reduced recurrent arch obstruction. Could this technique also contribute to the incidence of left-sided PVO? Table 1 from the article of Kotani and colleagues¹ indicates that 5 of the patients with PVO had hypoplastic left heart syndrome (all presumably underwent stage 1 palliation), and 5 other patients had diagnoses consistent with possible previous stage 1 palliation (tricuspid atresia with transposition, n = 3; double-inlet right ventricle, n = 2). Of these 10 patients, only 3 had successful palliation, and the outcomes for those with hypoplastic left heart syndrome were dismal. Two questions come to mind regarding the group of patients with previous stage 1 palliation that includes arch repair: does this group warrant early and more frequent surveillance with CMR to detect PVO? Could posterior descending aortopexy by left thoracotomy, either alone or in combination with a staged sutureless pulmonary vein repair, be more beneficial in the treatment of PVO? Of the anatomic factors implicated in the pathogenesis of left-sided PVO, it would seem that malposition of the descending aorta would be most easily modified with posterior descending aortopexy. From a brief literature search, there are no reports of successful relief of left-sided PVO apart from the cases included in this report with the sutureless repair. Kotani and colleagues¹ have not used posterior descending aortopexy; consequently, any therapeutic benefit is speculative at this time.

Finally, Kotani and colleagues¹ deserve sincere congratulations for another very valuable contribution to the management of one of the more vexing groups of patients who have PVO develop. Their efforts to elucidate an anatomic explanation for the development of left-sided PVO in patients with single-ventricle anatomy may well lead to effective ways to prevent or correct PVO that will increase the rate of successful palliation for this difficult subset of patients.

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