There Are Some Things Money Can Buy

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Some pathologies that are managed by pediatric cardiothoracic surgery have stalled in the attempt to improve the “natural history of intervention”; functional single ventricle with TAPVC is one such entity.

Central Picture Legend: Camille L. Hancock Friesen, M.D.
Geoffrion et al. have updated the Children’s Hospital of Philadelphia experience in patients with functional single ventricle (SV) and total anomalous pulmonary venous connection (TAPVC) which was first reported in 1999 \(^1\)\(^2\). This four-decade retrospective (1982-2022) single-center series includes 190 children with SV/TAPVC. The authors have divided the cases into three nearly equivalent eras, and have shown that early and late mortality rates plateaued after the first era, leaving this group with a 50% 10yr survival rate. It is worth bearing in mind that the definition of early mortality in this manuscript is different to that employed by the Society of Thoracic Surgeons. Early mortality in this series includes both operative and nonoperative patients and early mortality is limited to 30d post operatively (regardless of length of stay hospital). The authors have shown, unsurprisingly, that the highest risk subsets are patients with HLHS and those with obstructed TAPVC (HR for early mortality of 1.6 and 1.8 respectively). It bears remembering that BTS/TAPVC repair is THE operation with the highest STAT score of any procedure in the Society of Thoracic Surgeons Congenital Heart Surgery Database (STAT category 5, STAT score 5 mortality risk 38.7% (range 28.9%-49.1%)\(^3\). This is important context for young and emerging clinicians, cardiologists and cardiac surgeons alike, given our generally high (and usually appropriate) expectation of a good outcome in an era when overall program mortality rates are <3% (STS CHSD accessed 10/31/2023).

The authors have generously shared the following “pearls” about their programmatic approach to SV/TAPVC patients: 1. They have not used ductus venous stenting as a temporizing strategy although it has been described\(^4\). 2. They rarely (n=1) use Hybrid stage I palliation (PDA stent/bilateral PA bands/+/-atrial septectomy) in patients with unobstructed TAPVC although this is a strategy used for high-risk patients in some centers\(^5\). 3. They do not consider patients with PA/TAPVC a contraindication to surgical intervention whereas some centers would consider this a contraindication. 4. Only two of their 13 transplants were primary operations. 5. They convene a multidisciplinary ad hoc discussion around each patient and do not have a “protocol” per se for patients with SV/TAPVC.

In this heterogeneous group of patients (different types of functional single ventricle, different types TAPVC) 10y survival has not changed over the past 20y. Given the previous pathologic finding in all SV/TAPVC patients of dilated pulmonary veins with increased wall thickness and tendency to “arterialization”\(^6\)\(^7\) there may be some merit to conceptualizing the fate of the pulmonary venous system as the early dominant pathology in SV/TAPVC. When we look at the recent 5-year outcomes for recurrent pulmonary vein stenosis using an “anatomically-focused repair strategy” for pulmonary vein obstruction (PVO, primary or acquired; a series in which 30% of patients have single ventricle physiology)\(^5\) there has been a significant era improvement as a result of multidisciplinary team-based approach with specific vigilance for the status of each pulmonary vein and a lower threshold to intervene to reduce turbulent flow by whatever means. While the PVO and SV/TAPVC cohorts are not exactly analogous, it is notable that the 5y survival of the PVO cohort is 50%, the same as the 10y survival for the SV/TAPVC cohort. Perhaps we have done all we can to optimize the outcome in these patients, and precision medicine with cellular and subcellular targets will be the next phase of improved outcome.

There are some things that money can buy—but for everything else we, as a discipline, continue to carefully acquire and comb data to glean any learnings that will enhance outcomes for our patients and their families. Geoffrion et al. have provided important current benchmarks for a challenging cohort.
References


