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Commentary: Obstructed single ventricle total veins: Perhaps emergency surgery is not always the best answer

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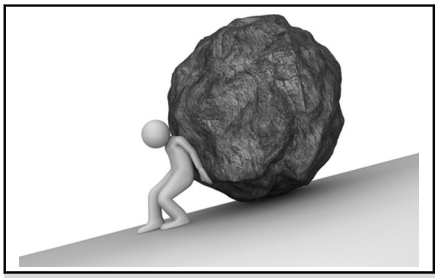

The Sisyphean task of emergency surgery for obstructed single ventricle total veins.

CENTRAL MESSAGE

Primary draining vein stenting followed by delayed surgical repair may be a superior management strategy for single ventricle patients with obstructed total anomalous pulmonary venous return.

The classic approach of emergency surgical repair for single-ventricle (SV) patients with total anomalous pulmonary venous return (TAPVR) and pulmonary venous obstruction at times appears like a Sisyphean task—laborious and generally unsuccessful.[1-5] The challenges of managing a balanced parallel circulation in the setting of pulmonary hypertension, and developmental and postbypass lung injury are often insurmountable. Given the lack of significant improvement in survival rates for this exceedingly challenging patient group, any alternative treatment approaches should be given serious consideration.

Kisamori and colleagues[6] report their outcomes with primary draining vein stenting (DVS) before definitive surgical repair in SV patients with obstructed TAPVR. In their small, single-institution cohort study, the authors found patients who received primary DVS followed by delayed surgical repair had significantly improved 90-day (100% vs 55.6%) and 5-year survival (54.9% vs 38.9%) compared with patients who received conventional primary surgical TAPVR repair.[6] After primary DVS, patients underwent surgical TAPVR repair at a median age of 88 days, compared with 8.5 days of age for patients undergoing primary repair. The only unique complication reported was postsurgical repair transaminitis that occurred in 3 of the 4 patients who underwent primary DVS of the ductus venosus, and resolved with stent coil embolization. These improved survival results lead to a compelling argument for a less-is-more approach during the neonatal period, and shifting the surgical repair until after a period of pulmonary venous decompression and lung recovery.

Although the authors present compelling data for consideration of primary DVS before definitive surgical TAPVR repair, the lack of randomization inevitably results in selection bias and the inability to establish superiority of this approach—a limitation the authors openly acknowledge. For instance, the degree of preoperative obstruction may have been more severe in the primary surgical repair group, as evidenced by earlier time of intervention, with 44.4% of surgical patients requiring intervention on day of life 1 compared with 18.2% of patients receiving primary DVS. Compounding the issue of selection bias is the small sample size, which is not unexpected given the rarity of this constellation of congenital cardiac defects.

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Despite the limitations, the authors present a convincing argument for re-examining the current paradigm for management of SV patients with obstructed TAPVR. The survival outcomes for patients who underwent primary DVS are impressive enough to warrant a closer look at this strategy, which allows for a period of lung recovery and transition out of the fragile neonatal period before the insults of definitive surgical repair. However, there are also significant challenges with this approach that will likely require ongoing refinement, including the technical aspects of stenting as well as the strategies for management of pulmonary blood flow before repair. Overall, the authors present an intriguing suggestion to shift the current treatment paradigm for SV patients with obstructed TAPVR from immediate to delayed surgical repair by way of DVS.

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