Optimal Management Strategies for Patients With Complex Congenital Heart Disease

Rae-Ellen W. Kavey, MD, MPH

The combination of critical pulmonary outflow obstruction with a large ventricular septal defect is symbolized by the diagnosis of tetralogy of Fallot, the most common form of cyanotic congenital heart disease. In many ways, the history of management for patients with tetralogy is representative of the evolution of management for all children with complex congenital cardiac diagnoses. Tetralogy was the first complex lesion addressed surgically with the palliative Blalock-Taussig shunt in 1945 and the first repaired completely with patients’ mothers serving as the bypass machine in 1954.1,2 The introduction of valved conduits in 1964 allowed repair of lesions not previously considered amenable to correction.3 The next 3 decades were marked by improved survival at open heart repair, recognition of the progressive deterioration of valved conduits necessitating early and repeated reoperation, and a progressive decrease in the age of primary repair to now the first few months of life.4,5 In the late 1970s, reports of arrhythmia and late sudden death among tetralogy survivors led to evaluation of tetralogy cohorts with residual right ventricular (RV) volume and/or pressure overload identified as predictors of late complications.6,7 Recognition of this problem heightened appreciation of the importance of arrhythmias in postoperative congenital heart disease patients and identified the need for informed long-term care and attention to residua of earlier procedures for all patients with congenital heart disease.8 Finally, the last 20 years have witnessed the dramatic rise in interventional procedures to address both correction of simple lesions and palliation/management of complex diagnoses.9

The remarkable progress in care of children with tetralogy exemplifies the collaboration between pediatric cardiologists and congenital cardiac surgeons, the development of new techniques to address problems as they emerge, and the continuous evolution of management that have characterized care of all patients with congenital heart defects in the last 60 years.10 Despite the remarkable progress made, tetralogy also symbolizes the difficulty of critically evaluating questions in patient management when the number of patients in any one center is relatively small and when patient management continues to evolve. Although publications on congenital heart disease are numerous, most are retrospective case series with few prospective and/or randomized approaches to evaluating management questions.11

The report by Peng et al12 in this issue of Circulation encompasses all these issues. The article reports the results in 221 patients who underwent placement of 242 balloon-expandable bare metal stents to address obstructed RV-pulmonary artery (PA) conduits, the majority in patients with complex forms of tetralogy of Fallot. This is the largest reported series of stent implantation to address conduit obstruction and confirms earlier reports in smaller numbers of patients that the technique can be applied successfully in this setting.13,14 Results immediately after stent placement indicate a significant decline in RV pressure, and the authors report a delay in the need for conduit replacement surgery averaging 3.9 years in children >5 years and 2.7 years overall. There were no deaths, and procedural morbidity was minimal. No systematic change in RV function was demonstrated. At catheterization, a mean of 2.2 years after the initial stent procedure, mean RV pressures had returned to preprocedural levels. An unexpected finding was the presence of significant stent fractures in 43% of those evaluated fluoroscopically; in most cases, there was major disruption of the stent integrity. Fractures were associated with compression of the stent and substernal location and were visible only with dynamic imaging; there were no identified hemodynamic consequences. The authors conclude that conduit stenting effectively addresses RV-PA obstruction and prolongs conduit lifespan.

Peng and colleagues12 are to be commended for extending the pediatric cardiology/congenital cardiac surgery tradition of developing innovative ways to address the problems associated with management of congenital lesions and the complications of previous interventions. Any alternative to a standard therapy must demonstrate a favorable balance between risk and benefit. Unfortunately, in this retrospective series, uniform evaluation of the results of stenting was not performed; thus, significant questions remain about applying the technique in this setting. Cardiac catheterizations in 57% of patients demonstrated that RV pressures had returned to preprocedural levels at a mean of 2.2 years after stent placement. Presumably, patients whose clinical or echocardiographic findings suggested increasing obstruction are those referred for repeated catheterization, so these findings may not be generalizable to the entire group, but it is possible that stenting offers only a very temporary decrease in RV outflow obstruction. Complete results of postprocedural echocardiograms are not reported; they would have been useful for evaluating the duration of the response to stenting.
and the impact of pulmonary insufficiency, a universal sequela of stent placement, on RV size and function. Stent fractures were discovered incidentally at catheterization or surgery; when did they develop and what will be their clinical consequences? How did the presence of the stent impact subsequent RV outflow tract surgery, something that occurred in 70% of the patients at a mean of 2.7 years after stent placement? With no standard protocol for serial evaluation of conduit gradients, RV size and function, and stent integrity, the ultimate benefit of a temporary decrease in RV pressure, especially when accompanied by the development or progression of pulmonary insufficiency and by the risk of stent fractures, is unclear.

A technical advance like the one described here represents both an option for management of the patient with RV-PA conduit stenosis and an opportunity to reevaluate the overall approach to care in this group of patients. All currently available conduit valves deteriorate, and the smaller the conduit at original repair, the earlier progressively more abnormal hemodynamics will develop. The reported mortality rate at repeated conduit surgery is 1.7% to 4.9%, and morbidity is high. Until now, the management strategy has been to accept significantly abnormal hemodynamics, often for many years, delaying the need for additional surgery as long as possible. However, the overall goal for patients like this has changed from the early days of intervention for children with tetralogy from survival alone to normalization of anatomy to the greatest extent possible with optimal preservation of cardiac function. When RV outflow tract stenosis develops after conduit insertion, what is the right time to intervene to reduce RV pressure when the objective is optimizing RV function? Does a decrease in pressure, especially when accompanied by an increase in RV volume overload, represent any net improvement in terms of maximizing RV performance? What are the sequelae, beyond the immediate risk of procedural mortality and morbidity, for repeated RV outflow tract stent placement with associated radiation exposure? Should magnetic resonance imaging rather than angiography be used as the imaging modality to address this risk? What are the functional sequelae of repeated surgical conduit valve replacement? If trading pressure normalization for a degree of volume overload is the right approach, then serial stenting to keep RV systolic pressure as low as possible should be the right course. In that setting, what are the potential late problems associated with stent fracture? If volume overload secondary to pulmonary insufficiency is the major culprit in the development of RV dysfunction, then early valve replacement should be the strategy. These are just a few of the questions raised by confirmation of the utility of this new procedure. Further advances like percutaneous pulmonary valve replacement and the development of tissue-engineered pulmonary valves seeded with a patient’s own cells make certainty of the best strategy for management even more important.

Even with a standard protocol, no single center can effectively address management questions like these because patient volumes with a single diagnosis are small. Over a 15-year period, the number of patients referred for management of obstructed RV-PA conduits to the very well-regarded interventional catheterization program that reported this series averaged just over 1 per month. Only a protocol-based multicenter comparison of surgical and catheter intervention with standardized serial evaluation of hemodynamics and function has the potential to critically evaluate short-term, mid-term, and long-term results. Findings from studies like the one by Peng et al will ultimately allow the development of evidence-based management strategies for complex congenital heart disease. In 2001, the National Heart, Lung, and Blood Institute established the Pediatric Heart Network, a new program to provide the infrastructure for multicenter studies. With this support, a collaborative group of pediatric cardiology/congenital cardiac surgery programs has been established and is beginning to amass critical volumes of systematic data from observational studies and trials. The Pediatric Heart Network is not the only forum for multicenter clinical research in pediatric heart disease, but it has already demonstrated the power of combined volumes, common infrastructure, and sustained collaboration. To half a century of outstanding technical progress we will now be able to add the results of equally expert evaluation protocols, powered to answer critical questions in patient care.

Management of congenital heart disease has come an enormous distance since Drs Blalock and Taussig first worked together. Cardiology and cardiac surgery teams cooperating in multicenter clinical research represent an extension of that philosophy; their findings should allow us to develop strategies that will offer our patients optimized anatomic repair, preserved cardiac function, and the best possible quality of life.

Disclosures

None.

References


**Key Words:** Editorials | heart defects, congenital | tetralogy of Fallot