

More Is Not Always Better, Less May Be!



James D. St. Louis, MD, and James E. O'Brien Jr, MD

The evolution of the surgical management of tetralogy of Fallot parallels the history of congenital heart disease in general. The first pathologic description of tetralogy of Fallot occurred in the 17th century.¹ It has been 75 years since initial operative palliations were successfully achieved, but it wasn't until the early 1980s, with the widespread use of cardiopulmonary bypass, that intracardiac correction of this defect became reproducible with excellent operative outcomes.^{2,3} Unfortunately, these successes have come with a price: an entire generation of patients with significant unanticipated long-term morbidities has been created. With our evolving understanding of these long-term sequelae, it has become evident that several of the specific technical maneuvers routinely applied have resulted in permanent and lasting anatomic lesions with significant limitation in physical well-being and quality of life. Two such residual lesions are the significant pulmonary regurgitation created when a hypoplastic pulmonary annulus is opened and the creation of an extended right ventricular incision to relieve outflow tract obstruction and better expose a malaligned ventricular septal defect.

In this edition of *Seminars in Thoracic and Cardiovascular Surgery*, Simon et al.⁴ present a single-center experience comparing 2 noncontemporaneous cohorts defined by the extent of the right infundibular incision created at the time of surgical repair. Thirty-eight patients over a 30-year period were retrospectively reviewed. The study concluded that limiting the length of the right ventricular incision was associated with improved early and late outcomes, conferring a 10-year advantage in freedom from reoperation, better cardiopulmonary fitness, and less right ventricular dilation. The authors should be applauded for providing statistically significant evidence that supports current surgical strategies limiting the extent of the transventricular incision, therefore optimizing not only successful short-term survival, but assuring improved long-term functional capacity as well.

It is truly astonishing to have had the privilege of witnessing the evolution of congenital heart surgery over such a short period of time. Already having a keen interest in congenital heart surgery and while a second-year medical student at Georgetown University, some 32 years ago, I remember being impressed by



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Central Message

Extended right ventriculotomy increases long-term morbidities associated with the surgical repair of tetralogy of Fallot.

watching a famous cardiac morphologist dissect several hearts from patients who had undergone successful operative correction of their heart defect, but later succumbed for noncardiac reasons. I recall the aggressive removal of a long autologous pericardial patch from the right ventricle, crossing the incised pulmonary annulus and concluding on the pulmonary artery. I remember the professor's authoritative comments convincingly professing that the severe pulmonary insufficiency created by the widely open pulmonary annulus and the extensive incision in the right ventricle would not result in any long-term consequences, emphasizing the limited importance of RV function on long-term survival and physical activity. Clearly, a lesson into our limited understanding of the long-term consequences of the successful, although aggressive application of the surgical techniques of the time. It would behoove our current generation to heed these mistakes of the past. Perhaps we need to concern ourselves with many of the aggressive although successful management strategies we currently employ for children who would have certainly perished just a decade ago. One such group that particularly stands are patients who undergo functional single-ventricle palliations who succumb following years of abnormal physiology and aggressive use of mechanical circulatory support and cardiac transplantation. Please don't misunderstand this statement—I certainly do not suggest we should limit the availability of these revolutionary technologies. But, we must have a firm grasp of the consequences of successful application of these therapies. Further, we must be prepared to both acknowledge and support the generation that will ultimately fail these “super palliations,” finding and providing the resources to apply new technologies.

Ward Family Heart Center, Children's Mercy Kansas City, Kansas City, Missouri

Conflicts of Interest: None.

Address reprint requests to James D. St. Louis, MD, Ward Family Heart Center, Children's Mercy Kansas City, 2401 Gillham Road, Kansas City, MO 64108. E-mail: jdstlouis@cmh.edu

DOI of original article: <http://dx.doi.org/10.1053/j.semtcvs.2019.01.006>.

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