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EDITORIAL COMMENT

On the Evolution of the Fontan Operation

From an Electrophysiologist's Perspective*

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The history of pediatric electrophysiology as a subspecialty is inextricably bound up with the development of surgery for complex congenital heart disease, and in particular, the treatment of the arrhythmias that result from these palliative and corrective operations. In a pediatric electrophysiology center, the most problematic of these post-operative arrhythmias are arguably the re-entrant atrial arrhythmias, which account for many hospitalizations and interventions. Of all the types of repairs that are performed, it is the Fontan repair that has over the years created the most trouble with respect to atrial arrhythmias (1).

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The Fontan procedure itself has undergone numerous changes and continues to evolve. Initially, the Fontan consisted mainly of a direct connection between the right atrium to the pulmonary artery. Results were disappointing, both from hemodynamic and arrhythmia standpoints, and it was recognized that a great deal of hydraulic energy is lost in the atriopulmonary connection, leading to right atrial dilation, low cardiac output, and often, pulmonary venous obstruction. The prominence of atrial flutter (also known as intra-atrial re-entry tachycardia) led to the hypothesis that if poor hemodynamics and right atrial stretch could be ameliorated, the incidence of atrial flutter might be lowered. Further evolution of the Fontan involved various forms of cavopulmonary connection, allowing for more efficient streaming of venous return to the pulmonary bed with less loss of energy (2), and elimination of the potential for significant right atrial dilation. One method of cavopulmonary connection involves the creation of a lateral tunnel (either intracardiac or extracardiac), and the other involves placement of an external conduit, essentially bypassing the right atrium entirely. Although, in general, centers have observed a lower incidence of atrial arrhythmias compared with the old-style atriopulmonary connection, atrial arrhythmias have not been eliminated. Simultaneously, the role of long atrial suture lines as barriers to impulse propagation that support the development of atrial flutter has become well understood as, for example, in the Senning and Mustard procedure (3). Therefore, it is not surprising that atrial arrhythmias have not been eliminated simply by normalizing atrial pressures. The 2 main forms of Fontan cavopulmonary connection differ significantly in that the intracardiac lateral tunnel Fontan requires quite a lot of atrial surgery, whereas with the external conduit, the surgical impact on the atrium is less extensive. Some have proposed that a rationale for favoring the external conduit Fontan over the lateral tunnel would be the limitation of surgical impact to the atrium and, consequently, an expectation of a lower atrial arrhythmia incidence (4).

In this issue of the *Journal*, Stephenson et al. (5) report their findings in the analysis of patients enrolled in the Pediatric Heart Network's Fontan Cross-Sectional study. A total of 520 pediatric patients were enrolled and evaluated, all of whom had 1 of the 3 types of Fontan operation, and this is the largest series reported to date for whom this type of data has been collected. Not unexpectedly, the incidence of atrial arrhythmias in this group was high, 7.3%, with the prevalence increasing with age. Interestingly from the electrophysiologist's point of view, the incidence of atrial arrhythmia was in direct relationship to the amount of atrial disease and/or surgical impact: 19% in the atriopulmonary connection (highest, as expected), 7% in the intracardiac lateral tunnel (atrial surgery without atrial dilation), 5% in the extracardiac lateral tunnel (less extensive atrial surgery), and only 2% in the external conduit Fontan (the least surgical impact on the atrium). One notes that the difference in incidence was not statistically significant between the intracardiac lateral tunnel and the extracardiac conduit, although the study was powered to detect a hazard ratio of 2.5 or higher, and insufficient time may have passed to demonstrate more substantial differences.

So where does this leave us? It is disappointing, but perhaps not altogether surprising, that one observes some atrial arrhythmias in patients with the external conduit Fontan. Although Stephenson et al. (5) have not provided us with additional details of the operations, for many indications, an external conduit Fontan still requires a large atriotomy (e.g., to perform atrial septectomy or plicate a valve). The Fontan is not a legacy operation like the Senning procedure for transposition, and with improved survival for the Norwood/Sano operation, more and more children will be coming to surgery for Fontan completion, and so the management of these arrhythmias will continue to occupy us. Speaking from the point of view of the person holding the ablation catheter, all forms of cavopulmonary

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connection are problematic as they limit access to the atrial mass via the caval veins. The external conduit is likely the most problematic in terms of access. Various approaches are available, including crossing a fenestration, puncturing the conduit itself (6), entering the roof of the atrium via the superior vena cava and left pulmonary artery (7), direct transthoracic entry into the heart (8), and approaching the atria in a retrograde fashion after crossing the aortic and atrioventricular valves. None of these approaches are easy, and they all either limit catheter maneuverability or have significant risks of complications.

Perhaps, in view of the fact that we have been unable to avoid atrial arrhythmias despite extensive evolution in the Fontan procedure, it is time to consider what might be done at the time of Fontan completion to prevent these arrhythmias, as suggested by Collins et al. (9) in their prospective trial of strategically placed atrial incisions to prevent the occurrence of flutter. Certainly, after 15+ years of experience with entrainment mapping and electroanatomic mapping of arrhythmia circuits in patients after the Fontan procedure (3,10), we should be able to come to a better understanding of the exact elements of surgical technique that create the substrate for atrial arrhythmia, and apply this knowledge to prevent this surgical complication in the future.

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Key Words: arrhythmia ■ congenital heart disease ■ Fontan ■ intra-atrial re-entrant tachycardia ■ prevalence.