JACC: CLINICAL ELECTROPHYSIOLOGY VOL. 4, NO. 10, 2018

PUBLISHED BY ELSEVIER

EDITORIAL COMMENT

Ventricular Pacing in Single-Ventricle Complex Congenital Heart Disease

How Hard it Is to Achieve the Ideal*

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The decision to perform cardiac resynchronization therapy (CRT) in adult patients with systolic heart failure is made with the expectation of improving left ventricular function based upon thousands of patient-years of evidence and the knowledge of key factors that indicate potential responders to treatment. A minimally invasive (transvenous) implant can be accomplished in approximately 95% of patients.

In children, decisions related to pacing are substantially more complex. Infants and young children with structurally normal hearts and a need for pacing, such as those with congenital complete heart block, will need placement of an epicardial single- or dual-chamber system, using a surgical approach through full or limited sternotomy and/or thoracotomy. Although not impossible, the placement of a transvenous system in a young child is contraindicated given vessel diameter and risk of occlusion over time as well difficulties in lead placement due to cardiac size and the need to account for rapid physical growth. Surgical placement of epicardial leads in infants and children without prior sternotomy typically means full availability of the anterior ventricular epicardial surface for choosing ideal pacing locations. In children with normal cardiac biventricular structure who have cardiomyopathy or other forms of heart failure, CRT may be attempted, again through an epicardial pacing system. In this situation, placement of the left ventricular epicardial pacing lead is accomplished on the underside of the heart with less visibility for choosing the ideal site but with availability of a fresh epicardium.

In children with complex congenital heart disease with single-ventricle physiology, decisions related to pacing are exponentially more difficult. These patients will require epicardial pacing systems for their lifetime as opposed to children with normal cardiac structure where the change to a transvenous system is a future expectation. In single ventricles, the placement of a transvenous ventricular lead is contraindicated given that the lead would be in the systemic circulation with a high risk of causing embolic stroke. Furthermore, following stage III palliation with completion of the Fontan operation, venous access to the ventricular endocardium is physically nearly impossible.

In congenital complete heart block in children with normal two-ventricle cardiac structure, it is well known that there is a small risk of cardiomyopathy following initiation of single-site epicardial pacing. Ideally, pacing leads are placed closer to the ventricular apex to minimize pacing-induced dyssynchrony. It can be assumed that children with normal cardiac structure and function may tolerate variations in pacing site. However, such an assumption cannot be made for children with single-ventricle physiology. In hypoplastic left heart syndrome, the left ventricle is prohibitively underdeveloped, and surgical palliation is performed to use the morphologic right ventricle to pump the systemic circulation for a lifetime. Given decades of experience with these patients, it is well known that failure of the right

*Editorials published in *JACC*: *Clinical Electrophysiology* reflect the views of the authors and do not necessarily represent the views of *JACC: Clinical Electrophysiology* or the American College of Cardiology.

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The author attests he is in compliance with human studies committees and animal welfare regulations of the author's institutions and Food and Drug Administration guidelines, including patient consent where appropriate. For more information, visit the *JACC: Clinical Electrophysiology* author instructions page.

ISSN 2405-500X/$36.00 https://doi.org/10.1016/j.jacep.2018.09.002

ventricle is an expectation over time, regardless of the presence of atrioventricular synchrony with an intact conduction system. The prospect of new methods to avert or delay the failure of the systemic ventricle is beyond inviting.

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The study by O'Leary et al. (1) in this issue of *JACC: Clinical Electrophysiology*, highlights the difficulties of caring for patients with palliated single ventricles and conduction abnormalities requiring pacing. Although the limitations of a retrospective, single-center series with a diverse disease population are manifested by the statistical results, the importance of answering the question is paramount. Given the overall vulnerability of the single ventricle to progressive mechanical failure, it is logical that the goals of ventricular pacing are, first, to maintain atrioventricular synchrony through dual-chamber pacing and, second, to optimize ventricular activation to avoid electrical dyssynchrony. Achieving the second goal may be Herculean in difficulty given that selection of an ideal epicardial pacing site is not readily accomplished. Palliated single-ventricle patients have undergone multiple open-chest procedures, with each subsequent procedure requiring more extensive dissection to reach the epicardial surface. Placement of a new epicardial lead on the anterior surface is difficult enough without also attempting placement of a posterior epicardial lead in a location that allows reasonable pacing thresholds.

The question is whether such effort to perform multisite pacing is needed. Although there were no significant differences between dual-site and single-site pacing in reaching the primary endpoint of death or transplantation, the study by O'Leary et al. (1) demonstrated that single-site pacing in single-ventricle patients may be associated with a decline in ventricular function, whereas dual-site pacing for resynchronization may be beneficial, at least to potentially delay a decline in function or to prevent it altogether. There were no differences in other important markers of single-ventricle "health" such as the degree of atrioventricular valve regurgitation. However, the lack of statistical differences may be a consequence of the size of this study.

Given many factors to adjust, the decision to proceed with dual-site pacing in this patient population is clearly not straightforward. Many questions remain unanswered, including: what are the optimal positions of dual-site pacing leads for a single left ventricle or for a single right ventricle? What is the optimal method for achieving synchronization: QRS duration, mechanical function, or cardiac output? Is a "good" single pacing site better than 2 "bad" pacing sites due to limited access to viable epicardium? Although this study does not answer these questions, the data are intriguing, and the results warrant careful consideration when one is deciding the type of pacing system to implant in the single-ventricle patient. Furthermore, although the indication for pacing in this study was high-grade atrioventricular block, a real but relatively infrequent occurrence in palliated single-ventricle patients, the question remains whether elective multisite, epicardial CRT could halt the progression to heart failure in many single-ventricle patients. This study provides further impetus for performing the necessary multicenter trials, to the best of our ability, to help achieve the ideal long-term outcome for our complex congenital heart disease patients.

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KEY WORDS Fontan procedure, heart block, resynchronization