Assessing Ventricular Scar in Tetralogy of Fallot

Gregory Webster, MD

The prevalence of repaired congenital heart disease is increasing, and late mortality is an important clinical concern. Patients with tetralogy of Fallot (TOF) contribute the largest fraction of subjects in many articles that address late mortality. Multicenter studies and single-center groups have mined their databases for retrospective analyses in patients with TOF. These studies have tried to determine which interventions can alter the long-term course of the disease and when these interventions should be performed. In 2013, we know more about the long-term outcomes in TOF than in any other form of severe congenital heart disease. We have both surgical and catheter-based valve approaches to alter the major long-term hemodynamic problems. It would seem that if we can assist anyone with severe adult congenital heart disease, in TOF we stand on the tallest shoulders and might see the farthest.

For these reasons, we would hope to have the best success at treating ventricular tachycardia (VT) in this population. Antiarrhythmic medications and defibrillator therapies are offered to those patients with the highest risk of life-threatening VT. Ablation serves as a useful adjunct therapy, either to prevent future arrhythmia or to reduce implantable cardioverter-defibrillator shocks. In this issue of Circulation: Arrhythmia and Electrophysiology, Moore et al report the results of gross and microscopic evaluation from 27 postmortem specimens with TOF. The authors’ goal was to describe the dimensions of the right ventricular muscular isthmuses and quantify the degree of fibrosis. These isthmuses are narrow bands of conducting muscle; they are anatomically defined by features of the ventricle and by postsurgical scar. These isthmuses help complete the conduction circuits that produce macroreentrant VT. Blocking conduction through anatomic isthmuses is critical for successful ablation of VT. The current article adds an additional layer of data to the mental road map that electrophysiologists use to maximize the chances of successful ablation.

In 1992, Downar et al first recognized that life-threatening VT in TOF was originating in the right ventricular outflow tract. In 2007, Zeppenfeld et al laid out a map for the postsurgical right ventricle in TOF, using substrate mapping and postmortem specimens to describe 4 isthmuses that could support VT. All of the postmortem specimens of Zeppenfeld et al had a muscular isthmus between the right ventricular outflow tract patch and the tricuspid annulus, which they labeled isthmus 1. Only 1 specimen had an isthmus between the ventricular septal defect (VSD) patch and the tricuspid annulus, which they labeled isthmus 4. The conclusions of Zeppenfeld et al focused on the high-yield nature of ablating isthmus 1 between the right ventricular outflow tract patch and the tricuspid annulus, but the authors showed that ablating a combination of the 4 isthmuses could maximize success on a patient-by-patient basis. Unfortunately, even with this road map, catheter ablation has not been uniformly successful in alleviating VT in TOF.

Moore and colleagues want to improve the road map by more accurately describing the measurements of the muscle in these 4 isthmuses. In particular, Moore et al focus on the muscle that Zeppenfeld defined as isthmus 1. In the article of Zeppenfeld et al, isthmus 1 is bordered on 1 side by the tricuspid annulus, which they labeled isthmus 1A, and the top half as the isthmus along the VSD patch and the ventriculotomy (isthmus 1B). They report that isthmus 1B, bordered by the VSD patch, is thinner and shorter than isthmus 1A, bordered by the tricuspid valve. They hypothesize that isthmus 1B may be a better target for ablation.

Unfortunately, the distinction between 1A and 1B may be easier to appreciate on a gross pathology specimen than in an electrophysiology laboratory. In many cases, it may be impossible to distinguish 1A from 1B in the electrophysiology laboratory. Both the tricuspid valve and the VSD patch give off low- or absent-voltage signals and 2-point discrimination is limited by the electric characteristics of the catheter bipole, the accuracy of the mapping and recording systems, and the skill of the operator.

Even conceptually, ensuring that the VSD-to-ventriculotomy isthmus (1B) is the correct location for ablation requires determining whether conduction exists between the tricuspid valve and the VSD patch (isthmus 4). Again, the data from the article of Moore et al are helpful here, reinforcing the Zeppenfeld et al finding that isthmus 4 is rarely present. However, we know that the native atrioventricular conduction tissue in TOF runs inferior and posterior to the VSD, thus deep ablations on the inferior rim of the VSD should be conducted cautiously. If the isthmus between the tricuspid valve and the
VSD is a potential target for ablation, it may be worth considering whether an alternate lesion set could be designed for the patient that minimizes the chances of heart block.

The data of Moore et al7 on the length and thickness of each isthmus may become a helpful reference in designing potential lesion sets. However, despite their useful description of macroreentry corridors, these data have important limitations. The subject population in this study does not match the typical population who enters the electrophysiology laboratory. In this study, ≈40% of patients were <6 years, another 40% were between 6 and 20 years, and only 5 patients (20%) were >20 years. The average heart weighed only 135 g, and the average postsurgical survival was 1 day. The authors also considered a smaller group of their specimens, consisting of the subjects who were ≥5 years. This smaller group was useful because its selection excluded measurements based on infant hearts, which currently have little relevance for postoperative VT ablation. Even so, the average age in this group was 10 years, and the average survival after surgery in this group was 2 days. Only 6 patients were alive a year after the operation. This group remains younger, smaller, and more freshly postsurgical than the patients who typically present for ablation. As hearts with TOF grow to adult size, the isthmus measurements described here may not grow symmetrically; by the time the patient is ready for ablation, the relative lengths and thicknesses may differ substantially from these data. In addition, many hearts in this article were preserved within days or weeks of surgical intervention. Therefore, the microscopic assay reflected the extent of fibrosis at or near the time of repair. The progression of fibrosis over time is under-represented in these data, and that progression is likely relevant to ablation.

Despite these limitations, useful similarities exist between this study and current ablation practice. For example, the mean year of surgery in this study was 1973. The patients who were repaired in this era have grown into patients 30 to 50 years old who currently present with VT. The isthmuses that Moore and colleagues7 describe in this article continue to be present in patients with TOF who present for ablation. Their meticulous measurements represent an important quantification of the anatomy in these patients.

Monomorphic macroreentrant VT does not propagate randomly through the heart in congenital heart disease. Successful ablation requires the best possible understanding of anatomic substrate. Moore et al7 offer another layer of anatomic data in hearts with TOF, which may improve ablation strategies. However, TOF is just 1 lesion in congenital heart disease. We know that patients with transposition of the great vessels, single ventricles, left ventricular outflow anomalies, Ebstein anomaly of the tricuspid valve, and atrioventricular septal defects are all at high risk for sudden death.10 In time, we may understand the pathways for VT in these lesions with the same level of quantitative analysis that Moore and colleagues lead us toward in this article.

Disclosures

None.

References

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