

Ablation of ventricular ectopy in complex congenital heart disease: The best of both worlds



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In this issue of *Heart Rhythm Case Reports*, Mori and colleagues¹ present a case in which multiform ventricular ectopy was successfully ablated from the remnant of outflow tract septum in an adolescent with complex congenital heart disease. As the authors demonstrate with multimodality imaging and hand illustration, the ectopic focus was within the bar of muscle above a large ventricular septal defect and between the paired outflows in this patient with heterotaxy syndrome, double-outlet right ventricle, and prior single-ventricle palliation including surgical amalgamation of the proximal aorta and pulmonary artery (the Damus-Kaye-Stansel operation) and total cavopulmonary connection (the extracardiac conduit Fontan operation).

A retrograde approach was insufficient owing to the Damus-Kaye-Stansel operation and the subvalvar stenosis present in 1 of the outflow tracts. Success was instead achieved via an antegrade approach by ablating the site of the earliest prepotential, which was located on 1 side of the outlet septum. Venous access to the heart was obtained by puncturing through native tissue below the prosthetic Fontan conduit. This excellent technique² could perhaps be described as “sub-conduit puncture” rather than “transcaval puncture” to avoid confusion with the “transcaval access” technique increasingly used for transcatheter aortic valve implantation³ (puncturing from inferior vena cava to abdominal aorta to bypass unfavorable femoral artery anatomy).

The case is notable for those challenges the authors did *not* encounter, as a result of the careful preparation that is critical to success in congenital electrophysiology procedures.⁴ Such preparation generally includes (1) comprehension of the native anatomy and potential implications such as anomalous location of the conduction system; (2) detailed understanding of the prior procedural history, including review of the operative notes and catheterization reports; (3) careful review of prior imaging and consideration of additional imaging that

may help with procedural planning; (4) integrating the understanding of native and surgical anatomy with that of the arrhythmia of interest to identify procedural approaches and consider whether advanced interventional techniques may be needed to access the target; and (5) assembling a team that, individually or in sum, brings the skills needed to perform the procedure safely and successfully. In this case, it would have been easy to commit to a retrograde approach that turned out to be unproductive in this particular patient, or to have spent excessive time on the difficult task of puncturing a prosthetic conduit, or to have reached the area of interest but not identified the complex local electrophysiology that explained the multiple distinct morphologies of ventricular ectopy and indicated the best target for ablation.

That final aspect, involving the concept of a protected intramuscular origin with preferential conduction and multiple exits,⁵ exemplifies how intricate concepts born from adult electrophysiology literature and practice can and will be increasingly applied to congenital cases given the burgeoning adult congenital population.^{4,6} It remains to be seen what care model(s) will allow the integration of knowledge and skills from diverse sources so that the “best of both worlds” can be available to this cohort of extremely complex patients.

Acknowledgment

The author thanks Cheyenne Beach, MD, for suggestions on the manuscript.

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Funding Sources: This research did not receive any specific grant from funding agencies in the public, commercial, or not-for-profit sectors. **Disclosures:** The authors declare this manuscript was published in the absence of financial or non-financial relationships that could be considered a conflict of interest. **Address reprint requests and correspondence:** Dr Jeffrey M. Vinocur, Department of Pediatrics, Yale University School of Medicine, 333 Cedar St, New Haven, CT 06520. E-mail address: jeffrey.vinocur@yale.edu.