Catheter Ablation of an Intra-Atrial Reentrant Tachycardia in a Young Adult Fontan Patient With Complex Palliated Congenital Heart Disease

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Figure. (A) Computed tomography of the heart. A, systemic venous atrium; HV, hepatic vein; V, functional ventricle. Blue arrow, cardiac conduit. (B) Baseline electrocardiogram (ECG) and intracardiac tracing obtained during tachycardia. Surface ECG leads II, aVF, and V1, catheter in SVC and mapping catheter in free wall of atrium. (C) Activation map during tachycardia. AV, atrioventricular valve; LVZ, low-voltage zone. White, earliest endocardial activation; purple, latest activation; gray, low-voltage zone (<0.5mV). Arrow, macro-reentry impulse with clockwise pattern rotating around conduit. (D) Cardiac anatomy and retrograde approach for catheter ablation. Blue line, catheter ablation during radiofrequency ablation; red arrow, reentrant circuit.

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A 21-year-old male patient with congenital heart disease (CHD) had twice undergone pulmonary artery banding when he was 5 years of age, and then had surgery for single ventricle palliation including bi-directional Glenn anastomosis and Fontan completion (lateral tunnel variant) at 11 and 13 years of age, respectively. After his last surgery, he started to suffer from atrial tachyarrhythmia that proved to be resistant to different anti-arrhythmic medications and usually recurred promptly after cardioversion. Therefore, the patient was referred for catheter ablation. The patient had heterotaxy syndrome with polysplenia and univentricular heart (Figure A), interrupted inferior vena cava withazygos vein continuation, 2 atrioventricular (AV) valves with congenital mitral stenosis and situs inversus. During catheter ablation, atrial electroanatomic mapping through the right femoral vein ascending to azygos vein and superior vena cava (SVC), and then descending to the conduit was attempted unsuccessfully. Under the circumstances, transseptal puncture was impossible during ablation. Thus, we manipulated the mapping catheter through the tricuspid valve retrograde to approach the atrium. Using the NavX system (St Jude Medical, St Paul, MN, USA), we performed activation mapping of the atrium during tachycardia, which showed a macro-reentry atrial tachycardia (cycle length=250 ms) with clockwise pattern rotating around the central obstacle (low-voltage zone), located on the free wall of the systemic venous atrium with intra-atrial conduit forming part of the reentrant circuit and obstacle (Figures B,C). We proceed to perform a linear ablation between the low-voltage zone (free wall of systemic venous atrium) and the tricuspid annulus, which resulted in termination of tachycardia (Figure D). At 1 year follow-up there were no recurrent arrhythmias.

Ablation of atrial arrhythmias in pediatric and adult CHD patients has been well described, including the need for retrograde access, the utility and application of 3-D mapping, and the description of arrhythmia substrates, such as areas of low voltage/scar, natural and surgically created barriers, and chamber enlargement contributing to slowing of conduction to promote reentry. But the most interesting and compelling features of the present case were the complex anatomy, the retrograde approach used to ablate this arrhythmia, and the absence of both sinus and AV node dysfunction, especially for a polysplenic patient. In the present case, surface electrocardiography demonstrated an atrial tachycardia associated with an extensive low-voltage zone observed during 3-D mapping. A macroreentrant circuit in the clockwise direction travels around a central obstacle (low-voltage zone). The presence of barriers to conduction both natural and scar related, may provide the substrate for reentry in the setting of an enlarged atrium with conduction slowing due to prior surgical intervention. In scar-related intra-atrial reentrant tachycardia such as in the present case, the efficacy of ablation is dependent on correct atrial mapping to clearly define pathways of reentry. Catheter ablation using a retrograde approach can successfully eliminate complex intra-atrial tachycardia using a 3-D mapping system in a patient with heterotaxy syndrome with polysplenia (given the constellation of findings of interrupted inferior vena cava, azygos continuation and single ventricle). The greatest challenge, however, after Fontan procedure is accessing the left atrium from the SVC. To the best of our knowledge, this is the first documented case of intra-atrial reentrant tachycardia with heterotaxy syndrome including polysplenia without sinus node and AV node dysfunction after catheter ablation.

Retrograde access can be used safely, patients can tolerate this access approach, and there is no compromise to AV valve structure or function. This case also clearly highlights the utility of retrograde access and that this should be considered more often as an available tool in complex adult CHD ablations, particularly in patients with certain anatomical substrates.

References