Clinical Spectrum of Wolff-Parkinson-White Syndrome in Ebstein Anomaly: Insights into Electrophysiology and Catheter Ablation

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Introduction: Ebstein anomaly is an uncommon congenital abnormality that involves the right-sided cardiac chambers and is primarily due to failure of delamination of the tricuspid valve. This results in inferior displacement of the tricuspid valve and tricuspid regurgitation (TR), which are associated with the degree of atrialization of the right ventricle (RV). A significant number of patients with Ebstein anomaly develop cardiac arrhythmias during their lifetime including supraventricular tachycardia (SVT), atrial fibrillation and flutter, and ventricular tachycardia. The most prevalent arrhythmias in patients with Ebstein anomaly were atrioventricular reentrant tachycardia (AVRT) due to Wolff-Parkinson-White (WPW) syndrome. Most of the accessory pathways (APs) were right-sided.

Methods: A 22-year-old man with Ebstein anomaly associated with severe TR and RV failure underwent tricuspid valve replacement (TVR) with a bileaflet mechanical valve at age 16 years. He had WPW syndrome consistent with right posterior AP with antidromic tachycardia before the TVR. Surgical ablation for WPW was not performed during the surgery. Owing to early recurrences of tachycardia, the patient underwent a first catheter ablation attempt during post-operative recovery; however, it failed to eliminate AP. Thereafter, he experienced increasing burden from recurrent refractory wide QRS complex tachycardia despite oral administration of amiodarone 200 mg twice a day and required emergency cardioversion to restore hemodynamic stability. Transesophageal echocardiography confirmed the presence of thrombus in the posterior leaflets of the mechanical valve.

Result: Once the venous accesses were completed, he was properly anticoagulated with intravenous infusion of heparin. The CS catheter placement was challenging because the CS ostium was blocked by the mechanical tricuspid valve. Only small 6-F catheters were used to cross the mechanical valve and the CS was successfully cannulated. The His bundle location was confirmed below the mechanical tricuspid valve. Mapping and ablation were performed using a 7-F open irrigation ablation catheter (CoolFlex; St Jude Medical, Minnetonka, MN, USA). Supra-tricuspid valve mapping confirmed that the AP was located below the mechanical valve. To map and ablate the AP, the ablation catheter was placed across the mechanical valve and oriented posteriorly, avoiding direct contact with the mechanical valve leaflets or ring. The AP potential was recorded 5–6 mm below the tricuspid valve ring at the atrialized RV and the ablation at the site using 35 W with an irrigation flow of 30 mL/min eliminated AP conduction.

Conclusion: In patients with Ebstein anomaly and WPW syndrome, 3-D high-density electroanatomical mapping and catheter ablation across the mechanical tricuspid valve are feasible and useful.