CASE REPORT

UNUSUAL LOCATION of THE AV NODE in a PATIENT with WOLF-PARKINSON-WHITE SYNDROME and PRIMUM ASD

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Catheter ablation in patients with congenital heart disease can be very challenging. The anatomy of the conduction system can vary significantly. In this article, we present the catheter ablation procedure in a patient with primum ASD, Wolff-Parkinson-White syndrome and unusually located AV node.

Key words: Catheter ablation, WPW, congenital heart disease, tachycardia

INTRODUCTION
Catheter ablation of arrhythmias can be challenging in patients with congenital heart disease (1). Besides the complexity of the arrhythmias seen in these patients, variations in the anatomy of the normal conduction system also contribute to that challenge. Arrhythmias following surgical repair of congenital heart defects are particularly more difficult to deal with (2). In this article, we present the catheter ablation procedure in a patient with primum ASD, Wolff-Parkinson-White syndrome and unusually located AV node.

CASE
A 16-year-old male patient with a history of primum atrial septal defect, cleft mitral valve and Wolff-Parkinson-White syndrome presented to our institution for consideration of catheter ablation. At the age of one, he underwent repair of his atrial septal defect and cleft mitral valve along with surgical ablation of his accessory pathway which was reported to be in the right septal region. He had temporary AV block following surgical ablation. AV node conduction returned back to normal in few days. His accessory pathway conduction also recurred in few weeks. He continued to have occasional episodes of supraventricular tachycardia (SVT) and persistent preexcitation. He has had a recent episode of syncope which was preceded by his typical tachycardia symptoms. He has complaints of dizziness which were interpreted as of neurocardiogenic origin based on the typical clinical description of the patient. Due to the presence of occasional nonsustained episodes of tachycardia and persistent preexcitation pattern, an electrophysiology study with catheter ablation procedure was planned. A 3-catheter electrophysiology study was performed. One of the catheters was placed in the typical anatomical His location. However, no His electrogram was identified even in the posterior locations. This was initially interpreted as His electrogram being masqueraded by the prominent ventricular preexcitation. Atrial pacing demonstrated antegrade AP conduction without any block down to 240 msec. One of the catheters was placed in the coronary sinus and earliest retrograde conduction during right ventricular apical pacing was demonstrated in the right septal region. Further mapping during ventricular pacing as well as during delta wave mapping in sinus rhythm localized accessory pathway to right postero septal area, slightly anterior to the coronary sinus ostium. Mapping in the proximal coronary sinus region failed to show any earlier activation. Single atrial extrastimulus protocol did not reveal any dual AV node physiology. There was no reentry or SVT. Isoproterenol infusion was started at 0.015 mic/kg/min. Single atrial extrastimulus protocol revealed single reentry beats without induction of SVT. Double atrial-extrastimulus protocol induced nonsustained SVT utilizing right postero septal pathway. Cycle length of SVT was 213 msec. Termination was with antegrade block and spontaneous. RF ablation in the earliest right postero septal ventricular electrogram area (-13 msec from the QRS) resulted in successful termination of
accessory pathway conduction in < 3.5 secs (Figure 1).

Accelerated junctional rhythm with 1:1 VA conduction noted during ablation. At 27 secs of ablation, 2:1 AV block noted and RFA was discontinued immediately. AV node conduction recovered in < 2 secs. No more RF lesions were given. Average temperature of that RF ablation was 57 degrees. No more preexcitation was seen after that lesion. Surprisingly, a very small His electrogram was noted at the distal electrodes of ablation catheter (Figure 2). Further mapping in the right posteroseptal region demonstrated presence of His electrograms at this unusually posterior location, in the right posteroseptal area (Figure 3,4). His electrograms were not identified in midseptal or anteroseptal regions. Postablation testing failed to induce any reentry or SVT. Patient’s 12-lead ECG did not reveal preexcitation and the only abnormality was left axis deviation of the QRS. After the observation and testing period of 45 minutes, there was still no evidence of accessory pathway conduction and the electrophysiology study was concluded.

Figure 1- Successful ablation of accessory pathway in < 3.5 secs.

Figure 3- AP and lateral views of the ablation location. Mapping catheter is at the ablation location. His catheter is pulled back to the area with the most prominent His electrograms in the right posteroseptal area.
DISCUSSION

This case demonstrates that AV node location can vary significantly in patients with primum type ASD. Patients with ostium primum ASD or AV canal defects tend to have left axis deviation. A previous histopathological study showed that these patients have posterior displacement of the AV node and the His bundle (3). The postero-inferior displacement of the left bundle branch seemed to be responsible for left axis deviation. In our patient, location of the AV node was at a very posterior location. Therefore in patients with AV canal defects and ostium primum ASDs, if there is persistent ventricular preexcitation and His electrograms are not able to be identified, then RF ablation should be performed very carefully. Ideally such patients should have catheter cryoablation where freeze-mapping can help prevent AV node injury, if accessory pathway is in the septal region of the tricuspid valve annulus (4).
REFERENCES