## CASE REPORT

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

## Volkan Tuzcu, Amy Shah

University of Cincinnati, Cincinnati Children's Hospital Medical Center

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

**Correspondence:** Volkan Tuzcu, MD Cincinnati Children's Hospital, University of Cincinnati Divison of Cardiology, 3333 Burnet Ave, Cincinnati, 45229, OH, USA Email: volkan.tuzcu@cchmc.org 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 posteroseptal 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 posteroseptal pathway. Cycle length of SVT was 213 msec. Termination was with antegrade block and spontaneous. RF ablation in the earliest right posteroseptal ventricular electrogram area (-13 msec from the QRS) resulted in successful termination of



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

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 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.



Figure 2- Note the small His electrograms at the ablation catheter immediately following RF ablation. His catheter is placed in usual anatomical anteroseptal location. No His electrograms are present at that location.

#### 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).



Figure 4- RAO/LAO views of the location of His catheter and the mapping catheter at the coronary sinus os following RF ablation. Note the unusually posterior location of His.

## REFERENCES

- 1- Kriebel T, Kroll M, Paul T. Radiofrequency catheter ablation therapy in the young: current status. Expert Rev Cardiovasc Ther 2003;1(3):421-37
- 2- Veldtman GR, Hartley A, Visram N et al. Radiofrequency applications in congenital heart disease. Expert Rev Cardiovasc Ther 2004;2(1):117-26
- 3- Ih S, Fukuda K, Okada R, Saitoh S. Histopathological correlation between the QRS axis and disposition of the atrioventricular conduction system in common atrioventricular orifice and in its related anomalies. Jpn Circ J 1983;47(12): 1368-76
- 4- Gaita F, Riccardi R, Hocini M et al. Safety and efficacy of cryoablation of accessory pathways adjacent to the normal conduction system. J Cardiovasc Electrophysiol 2003;14: 825-829