Fasciculoventricular accessory pathways following repair of ventricular septal defects

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Introduction

The most common form of preexcitation in the pediatric population is due to Wolff-Parkinson-White syndrome (WPW) where atrioventricular (AV) conduction occurs partially or entirely through a congenital accessory AV bypass tract.[1] Acquired WPW has been described when AV accessory pathways develop across suture lines after surgery for congenital heart disease (CHD).[2–3] Fasciculoventricular (FV) pathways that manifest after CHD surgery have not been described previously. Although FV pathways do not participate in arrhythmias, their effects manifest as de novo preexcitation on surface electrocardiograms (ECGs).[4] We present 2 cases of apparent acquired FV pathways and describe the potential mechanism of development following CHD surgery.

Case 1

Background

A16-year-old patient who had previously undergone a surgical repair of a conoventricular ventricular septal defect (VSD) and subaortic membrane resection was referred for evaluation of ventricular preexcitation. The surgical repair had been performed when the patient was 4 years of age and had been complicated by postoperative Mobitz II and third-degree AV block with a 15-second pause, prompting the implantation of a permanent epicardial ventricular pacemaker.

Postoperative follow-up over the ensuing 8 years using periodic ECG, Holter recording, exercise stress testing, and pacemaker interrogation revealed the return of normal intrinsic AV conduction. Given these findings, the pacemaker generator was removed. On a follow-up evaluation, ventricular preexcitation was noted on an ECG. Further review of previous ECGs demonstrated no evidence of preexcitation before the initial reparative cardiac surgery but presence of it within 1 month after (Figures 1A and 1B). The patient never experienced palpitations, and supraventricular tachycardia (SVT) was never documented. Given the patient's history of postsurgical heart block, the decision to remove a permanent pacemaker, and the unusual ECG pattern observed, a diagnostic electrophysiology study (EPS) was performed to further evaluate the conduction system and define the etiology of preexcitation.

EPS description

A comprehensive EPS was performed at baseline and with isoproterenol infusion using standard diagnostic catheters in the coronary sinus, His bundle, and right ventricular (RV) apical positions. During sinus rhythm, the cycle length (CL) was 938 milliseconds, the AH interval was 90 milliseconds, and the HV interval was 30 milliseconds with ventricular preexcitation. Following the administration of 9 mg of intravenous (IV) adenosine, AH prolongation was noted without change in the HV interval or degree of preexcitation. A 12-mg dose of adenosine resulted in supra-Hisian AV block. RV pacing demonstrated the absence of retrograde ventriculoatrial (VA) conduction.

Atrial ramp pacing demonstrated AH prolongation without change in HV interval. At a paced CL of 480 milliseconds, abrupt loss of preexcitation occurred with HV prolongation to 70 milliseconds (Figure 2A). Atrial extrastimulus pacing resulted in progressive AH prolongation with fixed HV duration and preexcited QRS appearance. At

KEYWORDS Preexcitation; Congenital heart disease; Pediatrics; Ablation

ABBREVIATIONS **AV** = atrioventricular; **AVNERP** = atrioventricular node effective refractory period; **CHD** = congenital heart disease; **CL** = cycle length; **ECG** = electrocardiogram; **EPS** = electrophysiology study; **ERP** = effective refractory period; **FV** = fasciculoventricular; **FV-ERP** = fasciculoventricular effective refractory period; **RV** = right ventricular; **SVT** = supraventricular tachycardia; **VA** = ventriculoatrial; **VSD** = ventricular septal defect; **WPW** = Wolff-Parkinson-White syndrome

(Heart Rhythm Case Reports 2015;1:331–336)

Conflicts of interest: The authors declare no conflicts of interest.

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http://dx.doi.org/10.1016/j.hrcr.2015.05.005

KEY TEACHING POINTS

- In addition to postoperative heart block, electrophysiologists should also be aware of the possibility of the development of ventricular preexcitation after congenital heart disease surgical repair near the conduction system. Postsurgical ventricular preexcitation could result from injury to and recovery of conduction tissue near the site of surgical intervention.
- Postsurgical ventricular preexcitation has been associated with supraventricular tachycardia and can be treated with catheter ablation. In the setting of postsurgical preexcitation, careful assessment during electrophysiological testing is critical to determine the type of connection causing preexcitation and necessity for ablative therapy.
- Fasciculoventricular connections are an infrequently encountered cause of ventricular preexcitation. These connections have a fixed preexcitation pattern that does not change with atrial stimulation. Normal antegrade (and retrograde, when present) atrioventricular nodal conduction is observed during standard electrophysiological testing without inducibility of tachyarrhythmias. These connections have never been shown to cause tachyarrhythmias, and ablation of these pathways should not be performed.

S1/S2 600 milliseconds/430 milliseconds, abrupt HV prolongation and loss of preexcitation were noted (Figures 2B and 2C) followed by infra-Hisian AV block at S1/S2 600 milliseconds/340 milliseconds. These findings were consistent with an FV pathway with FV pathway block at 480 milliseconds and FV pathway effective refractory period (FV-ERP) of 430 milliseconds during an atrial-paced CL of 600 milliseconds. SVT was not induced with standard stimulation protocols. Catheter ablation was not performed on the bystander FV pathway.

Case 2

Background

A 19-year-old patient with a history of surgically repaired conoventricular VSD was found to have ventricular preexcitation. When the patient was 3 months old, VSD repair with patch closure had been performed. Preexcitation was not present prior to surgery. A postoperative ECG showed a right bundle branch block with left axis deviation consistent with bifascicular block and no preexcitation.

Follow-up ECGs obtained at subsequent clinic visits revealed 2 different QRS patterns. The first pattern demonstrated bifascicular block, whereas the second pattern showed preexcitation.

During adolescence, the patient began experiencing episodes of palpitations and lightheadedness. Comprehensive evaluations during episodes, including ambulatory rhythm recording, exercise stress testing, and tilt-table testing, revealed normal heart rate and blood pressure and no arrhythmias. Both QRS patterns were captured on a single ECG tracing during tilt-table testing (Figure 1C). Because of persistent symptoms and the abnormal ECG findings, an EPS was performed as a final diagnostic measure.

EPS Description

Standard EPS was performed in a fashion similar to that described for case 1. During sinus rhythm, the baseline CL was 1150 milliseconds, AH interval 80 milliseconds, and HV interval 26 milliseconds. In the preexcited state, the earliest RV activation was seen at the distal His bundle electrode pair. The AV node Wenckebach CL and AV node ERP (AVNERP) are demonstrated in Figures 3A and 3B. Of note, AVNERP occurred at 700 milliseconds/570 milliseconds before loss of preexcitation was observed. No changes in degree of preexcitation or HV duration were observed with pacing. Concentric decremental retrograde VA conduction was observed during ventricular pacing. Administration of IV adenosine in sinus rhythm demonstrated AV block without a preceding change in preexcitation (Figure 3C) and VA block with RV pacing. No arrhythmias were induced. These findings were consistent with an FV pathway that had block and ERP values equal to or shorter than those of the AV node. Catheter ablation was not performed, given a lack of associated significant arrhythmias.

Discussion

These 2 cases highlight the interesting finding of what appears to be acquired ventricular preexcitation following surgery to correct CHD, and, more specifically, VSDs. Postsurgically acquired preexcitation in the form of WPW has been reported in patients after the Fontan procedure, with accessory pathways located at the surgical anastomosis of the right atrium to the right ventricle.[2–3] These pathways have been reported to have typical properties of WPW, facilitating orthodromic reentrant tachycardia and rapid AV conduction of atrial arrhythmias. To the best of our knowledge, this is the first case report describing postsurgically acquired preexcitation due to FV pathways.

In addition to WPW syndrome, preexcitation variants include atriofascicular, nodofascicular, nodoventricular, and FV pathways.[5] The FV pathway is characterized by a fixed pattern and degree of preexcitation, unchanged with rapid atrial pacing and atrial extrastimuli. It is the rarest variant of preexcitation, with a reported incidence of 1.2–4% in preexcitation cases, and it may even go unrecognized.[6–7] The pathway appears to arise from the His bundle or the bundle branches and inserts into the ventricular septum.[8] Decremental conduction has been reported but is generally

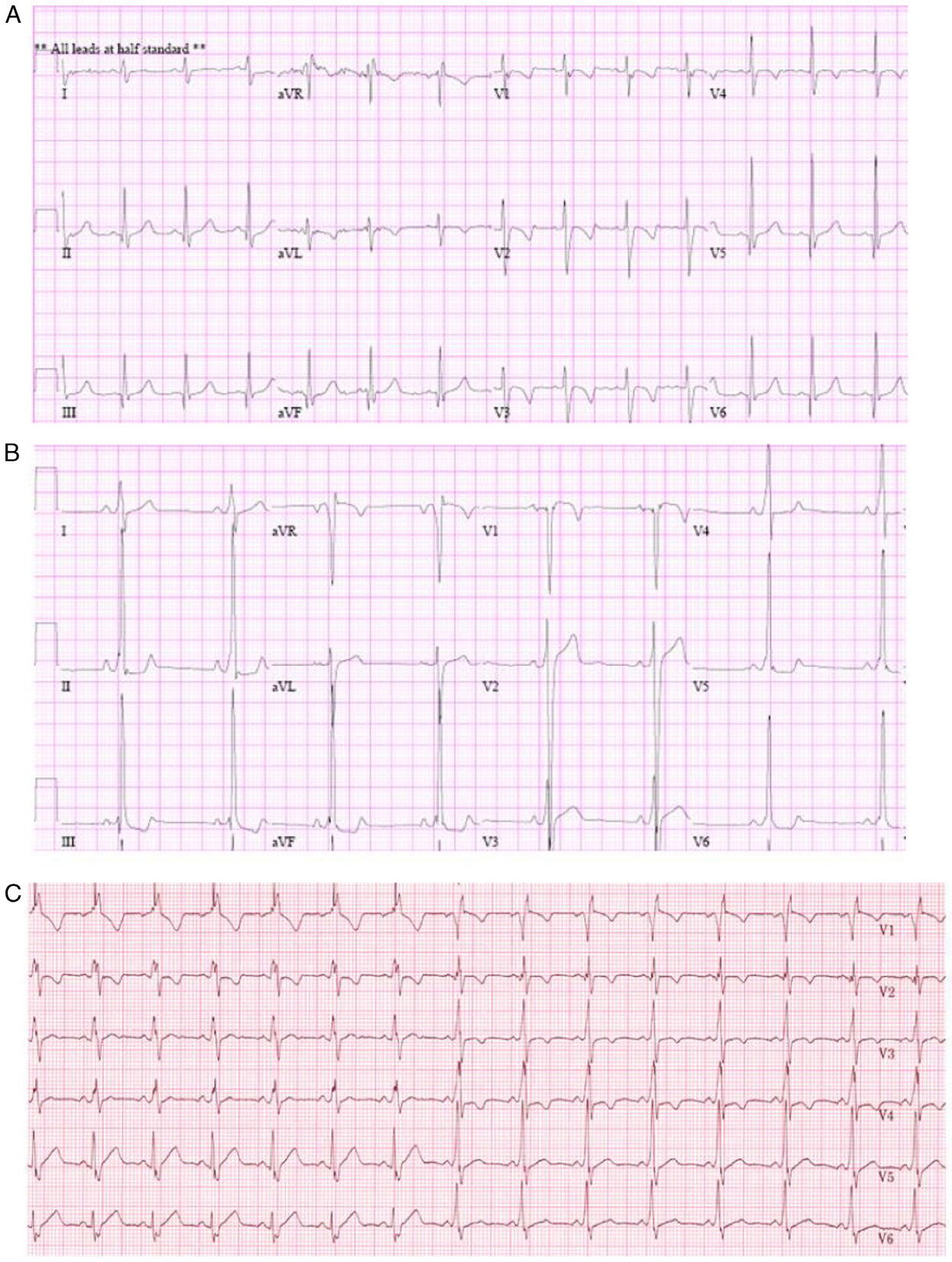

Figure 1 Cases 1 and 2 electrocardiogram (ECG) tracings. **A:** Case 1: Presurgery ECG demonstrating no preexcitation. **B:** Case 1: Pre-electrophysiological study ECG that demonstrates preexcitation. **C:** Case 2: Tilt-table ECG tracing capturing preexcited and nonpreexcited QRS complexes.

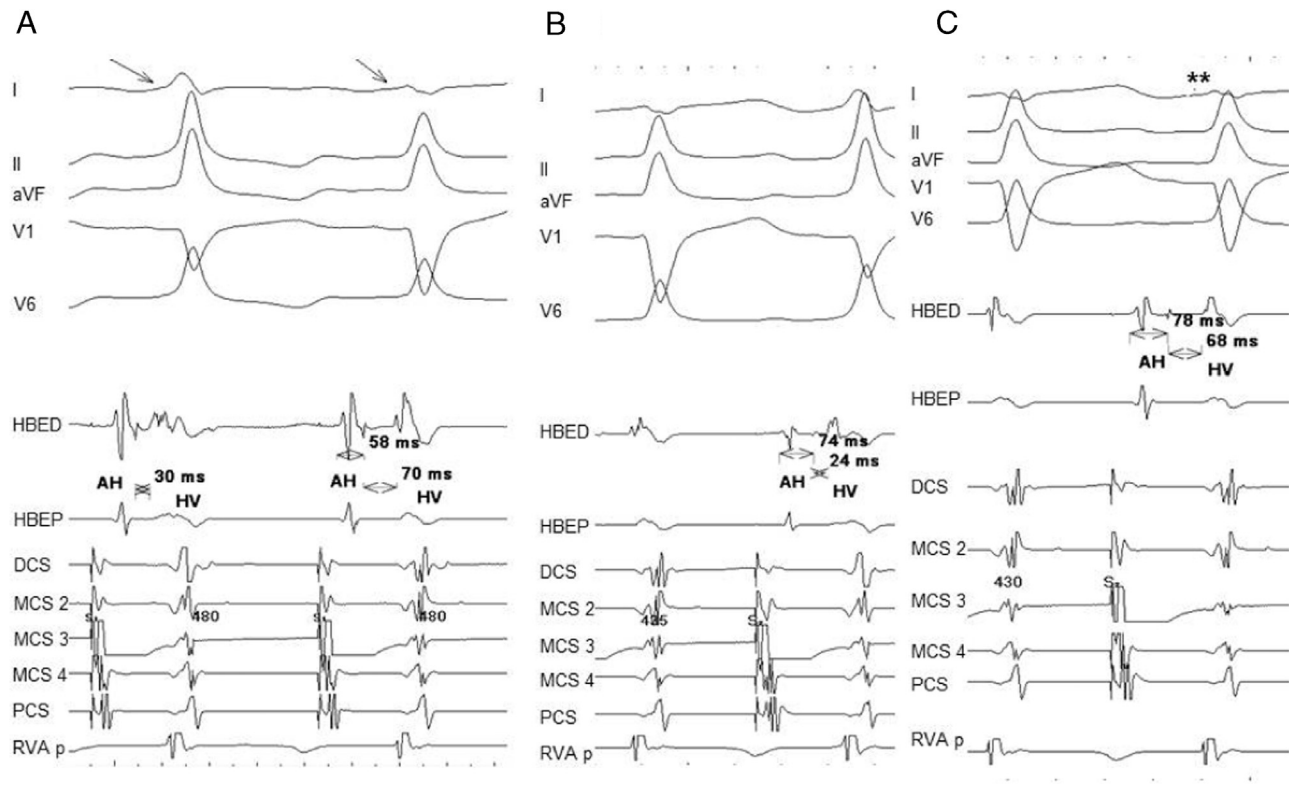

Figure 2 Case 1: Response to atrial pacing. **A:** With ramp pacing, abrupt loss of preexcitation occurs (arrows) at a paced CL of 480 milliseconds with prolongation of the HV interval from 30 to 70 milliseconds. **B:** With extrastimulus pacing, the degree and pattern of ventricular preexcitation was fixed during S2 conduction. Of note, preexcitation also abruptly disappeared and reappeared during the S1 drive. **C:** At S1/S2 600 milliseconds/430 milliseconds, loss of preexcitation occurs along with HV prolongation from 24 to 68 milliseconds (**), indicating FV pathway ERP and conduction through the His-Purkinje system.

uncommon.[9] FV pathways are mechanistically not involved in any type of tachycardia but may coexist as bystanders in association with rapidly conducting AV bypass tracts and during SVT. Therefore, in symptomatic patients, the etiology may be related to other arrhythmogenic substrates and not the FV pathway itself. The FV pathway can have overlapping ECG features with manifest anteroseptal and midseptal accessory pathways and therefore should be correctly identified to avoid unnecessary catheter ablation and inadvertent AV nodal damage.[4] Formal electrophysiological testing in our 2 cases revealed that the observed preexcitation patterns were consistent with FV connections rather than WPW syndrome. In both cases, there was no inducible SVT and the FV-ERP values were relatively long.

The specific mechanism for the development of FV connections in the 2 cases presented is not entirely clear. In conoventricular VSDs, the conduction tissue runs along the posteroinferior margin of the defect. Despite careful attempts to avoid conduction tissue damage, 1–4% of patients will require permanent pacing after surgical repair because of high-grade AV block.[10] A recent publication has shown that later recovery of AV conduction can occur after chronic postsurgical heart block.[11] The exact cellular mechanism of recovery is unknown, but a similar process may be responsible for the creation of a tissue substrate that permits preexcitation, as observed in the cases presented here. Tissue healing may have resulted in an electrical conducting bridge, similar to de novo atrioventricular bypass tracts seen after the Fontan-Björk procedure.[2] The growth of myocardial cells across the suture line or the presence of electrotonic transmission through this line are possible explanations for this finding. Further evidence supporting the potential for the growth of tissue across suture lines includes atrioatrial suture line conduction following orthotopic heart transplantation.[12] Another speculation into the mechanism of preexcitation in the cases presented here is that VSD repair resulted in disruption of the His-Purkinje fiber insulation, permitting direct connections from the His bundle or proximal fascicle directly to adjacent ventricular myocardium, resulting in an "acquired" FV pathway. Finally, it is possible that an FV pathway was preexisting but quiescent and was later "unmasked" following surgical damage to the native conduction system. Alternatively, if an FV pathway was preexisting in our 2 cases, the observed late manifestation may simply reflect age- and time-related changes similar to what has been described in other accessory pathways, such as WPW.[13–15] Direct histologic evaluation would be required to determine the specific etiology for the "new" finding of preexcitation in each of the cases presented.

Given the paucity of electrophysiological data on preexcitation emerging after CHD surgery, the decision to perform an EPS should be based on clinical judgment. Arrhythmias were not inducible in our 2 patients, and the FV-ERPs were relatively long, thus differentiating them from rapidly conducting AV bypass tracts.

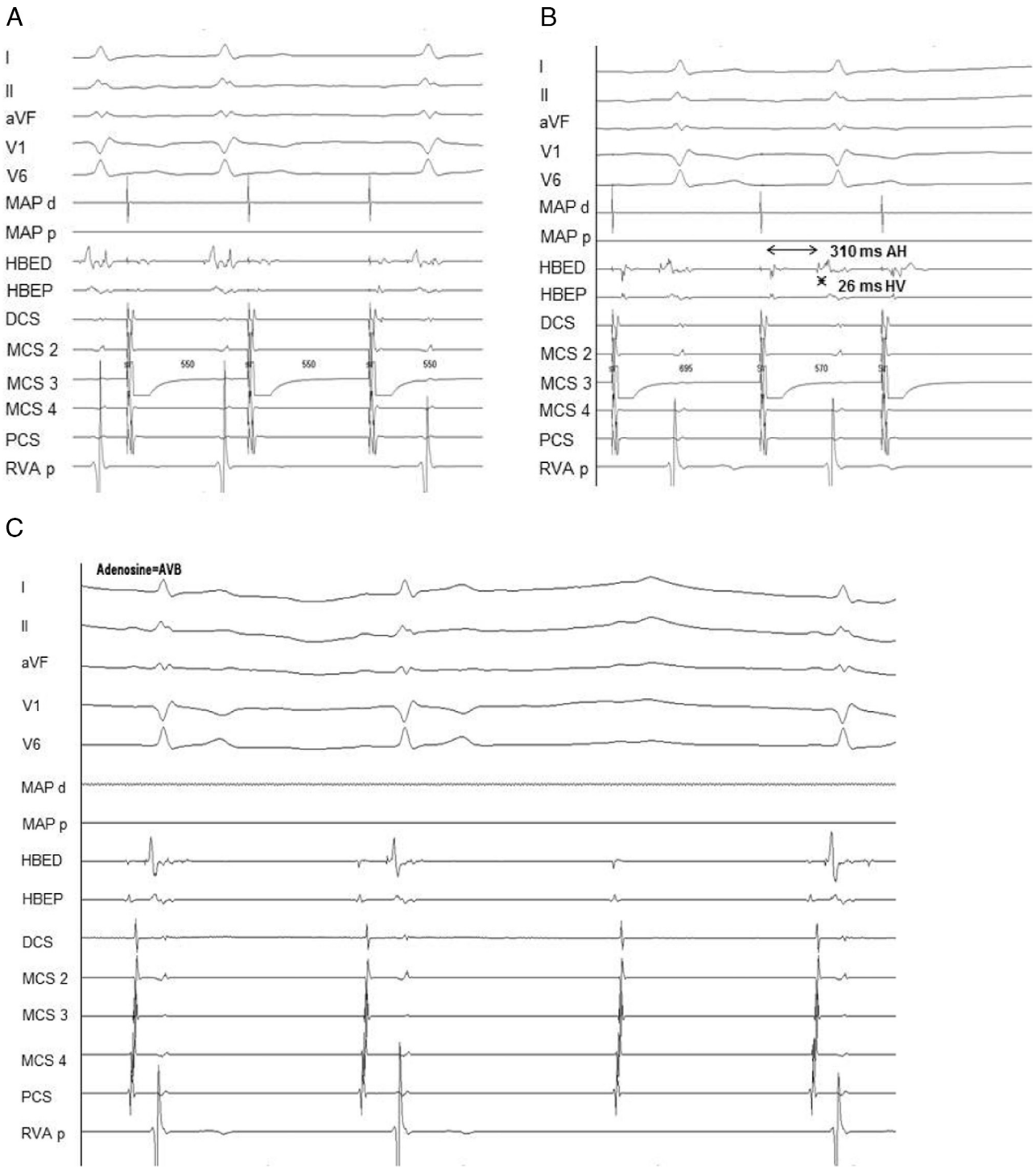

Figure 3 Case 2: Atrial pacing and adenosine administration. **A:** Whereas pacing at 550 milliseconds, AV nodal Wenckebach occurs without change in HV interval or QRS configuration. **B:** With extrastimulus pacing at S1/S2 700milliseconds/570 milliseconds, AVNERP is observed. **C:** Following IV adenosine, complete AV block occurs without change in preexcitation before or after block.

Conclusion

The finding of new postoperative ventricular preexcitation, an exceedingly rare finding, should raise the suspicion of an acquired accessory pathway connection. An invasive EPS can be performed to localize such a connection, assess its functional properties, and determine the presence or absence of clinically relevant arrhythmias.

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