# 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.<sup>1</sup> Acquired WPW has been described when AV accessory pathways develop across suture lines after surgery for congenital heart disease (CHD).<sup>2–3</sup> 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).<sup>4</sup> 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

**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. Address reprint requests and correspondence: Dr Philip M. Chang, Keck School of Medicine, Keck Medical Center of USC, 1510 San Pablo Street, Suite 322, Los Angeles, CA 90033. E-mail address: philip. chang@med.usc.edu. 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

## **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.<sup>2–3</sup> 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.<sup>5</sup> 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.<sup>6–7</sup> The pathway appears to arise from the His bundle or the bundle branches and inserts into the ventricular septum.<sup>8</sup> Decremental conduction has been reported but is generally

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