An acquired manifestation of fasciculoventricular pathway following complex congenital heart disease repair

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Introduction

Common forms of supraventricular tachycardia (SVT) are well characterized and include atrioventricular nodal reentrant tachycardia (AVNRT), atrioventricular reentrant tachycardia (AVRT), and atrial tachycardia. In pediatric populations, AVRT is relatively more frequent compared with AVNRT, in contrast with adult populations in whom AVNRT is the most commonly encountered subtype of SVT. The primary cause of AVRT is typically congenital, resulting from the failure of regression in atrial and ventricular connections, which supports the reentry mechanism. However, there have been occasional reports of acquired AVRT in patients with congenital heart disease following surgical repair.[1] In our case, we discussed the presence of fasciculoventricular (FV) pathways following the surgery.

Case report

A 9-year-old patient with a significant medical history of dextro-transposition of the great arteries, status post-arterial switch operation, including patent foramen ovale, ventricular septal defect (VSD) closures, and patent ductus arteriosus ligation since birth, was referred for evaluation of manifest preexcitation. The preexcitation pattern was not present before surgery (Figure 1A). Nine years later during follow-up, a repeated electrocardiogram (ECG; Figure 1B) and a 24-hour Holter monitor confirmed the presence of the preexcitation pattern. In addition, an ECG performed during follow-up showed sinus rhythm with delta waves, as illustrated in Figure 1B. There were no prior episodes of palpitations, syncope, or documentation of SVT in earlier ECGs. Given the patient's complex congenital heart disease, an electrophysiologic study was recommended for risk stratification.

KEYWORDS Acquired; Accessory pathway; Congenital heart disease
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KEY TEACHING POINTS

- Fasciculoventricular (FV) pathways represent a rare cause of ventricular pre-excitation in the general population. It is a congenital anomaly resulting from a disruption in the insulation of the His bundles/fascicles.
- The acquired form has been rarely described in the literature. Consistent with previous reports, our case study asserted the presence of FV pathway following complex cardiac surgeries for congenital heart disease.
- An electrophysiology study remains the primary diagnostic tool, supplemented by a thorough and detailed patient history.

Electrophysiologic study description

Per our institutional protocol, the electrophysiology (EP) study in pediatric patients was conducted under general anesthesia. Following the placement of venous sheaths on bilateral groins for the His bundle, coronary sinus, and right ventricle bipolar recording catheters, baseline parameters were obtained. The patient's baseline rhythm was sinus rhythm with a clear preexcitation pattern and a cycle length of 1085 ms. The AH and HV intervals were measured at 69 ms and 31 ms, respectively. Notably, V signal at HIS was earlier than V from RV apex catheters (Figure 2A).

Ventricular decremental pacing demonstrated concentric retrograde conduction, and a single ventricular extrastimulation test confirmed retrograde conduction via the AV node, evidenced by an increase in the VA interval, with the presence of a retrograde right bundle branch block. Atrial decremental pacing and atrial extrastimulation resulted in prolongation of the AH interval while the HV interval remained unchanged at 31 ms (Figure 2B). The degree of pre-excitation and QRS morphology also remained unchanged.

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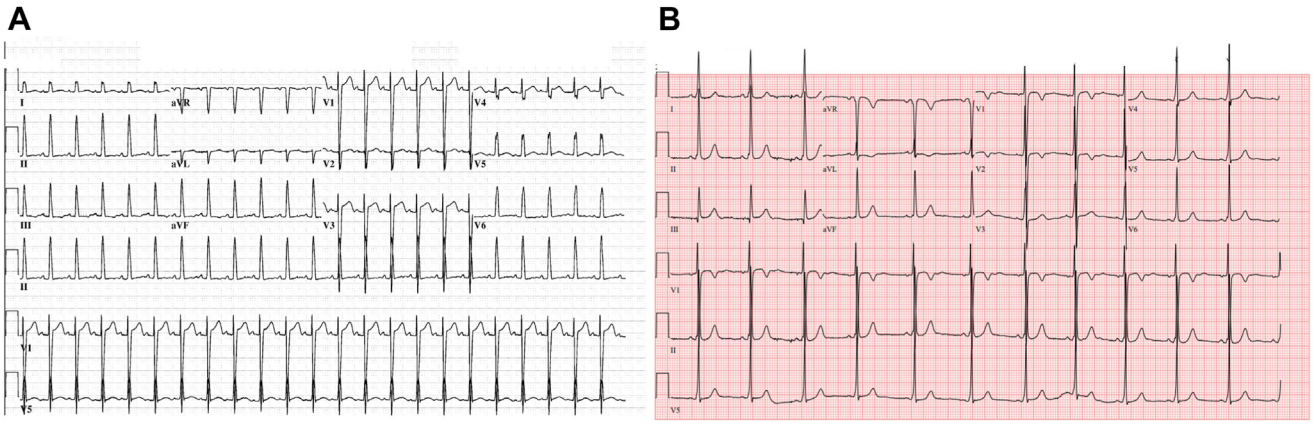

Figure 1 **A:** Preoperative electrocardiogram (ECG) prior to the patient's complex congenital heart surgery. **B:** Follow-up ECG after the index surgery.

The patient exhibited dual AV nodal physiology, indicated by an AH jump with rare single-echo beats.

For a comprehensive study, isoproterenol was administered and titrated to 0.04 mcg/kg/min. Repeated ventricular decremental pacing continued to show only concentric retrograde conduction. Ventricular extrastimulation protocols, with single and double extrastimuli, failed to induce any arrhythmias. Atrial burst pacing also did not result in any inducible arrhythmias. Notably, after atrial burst pacing, there was a sinus pause followed by a junctional escape beat, maintaining the same degree of preexcitation, QRS morphology, and HV interval as the baseline sinus rhythm. Next, adenosine was given which, again, only AH interval was prolonging while HV interval remained the same, at 31 ms (Figure 3A and 3B).

Based on these findings, it was concluded that the observed pattern is consistent with FV pathway. Given the well-described benign clinical course and the absence of inducible arrhythmias, no ablation was performed.

Clinical course

After the procedure, the patient has been doing well and reports no episodes of arrhythmia. Because of her progressively severe aortic stenosis after the arterial switch operation, she experiences only exertional dyspnea. She has been scheduled for regular follow-up with the pediatric cardiology team until she reaches an appropriate age for aortic valve replacement. Serial ECG monitoring has shown no changes.

Discussion

In our case study, we described an accessory pathway that can be acquired postsurgically, especially the FV pathway. In line with a prior report,[1] it is reasonable to conjecture that the VSD repair is most likely relevant to the pathogenesis in our case. We did not observe high-risk features of the accessory pathway in this patient, which aligns with the established understanding of the "benign" spectrum of this condition.[2]

In most circumstances, accessory pathways arise from abnormal tissue regression. During cardiac embryogenesis, only specific areas of cardiac tissue remain contiguous, which later form the sole route connecting the atrium and ventricle, AV node, and His bundle. Accessory pathways develop when additional areas fail to disconnect, potentially becoming arrhythmogenic substrates for supraventricular

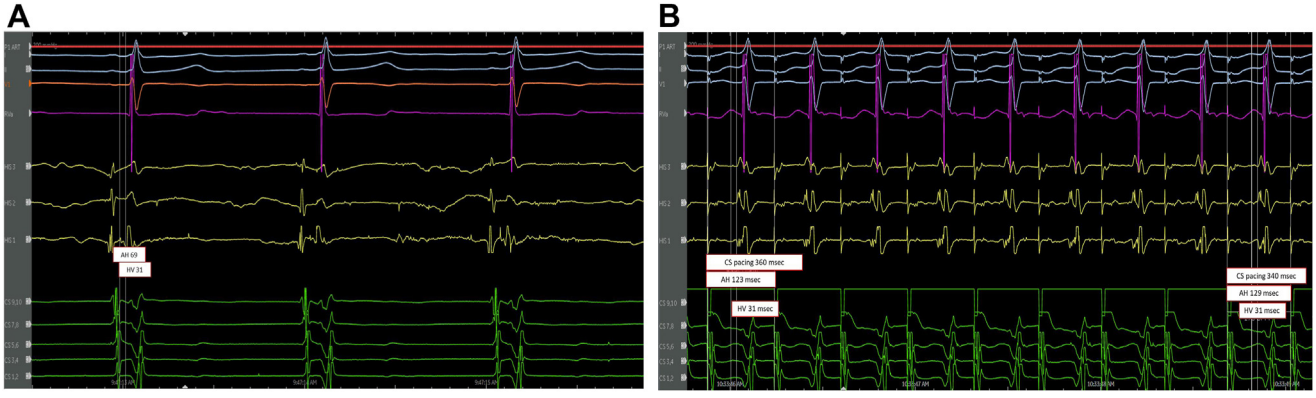

Figure 2 **A:** Baseline AH and HV durations are 69 and 31 ms, respectively. **B:** A slight prolongation on the AH interval, but stable HV interval, on atrial decremental pacing from proximal CS pacing at 360 and 340 ms. Of note, the V signal in HIS is earlier than in the RV apex. AH = atrium-his; CS = coronary sinus; HV = his-ventricle; RV = right ventricle; V = ventricle.

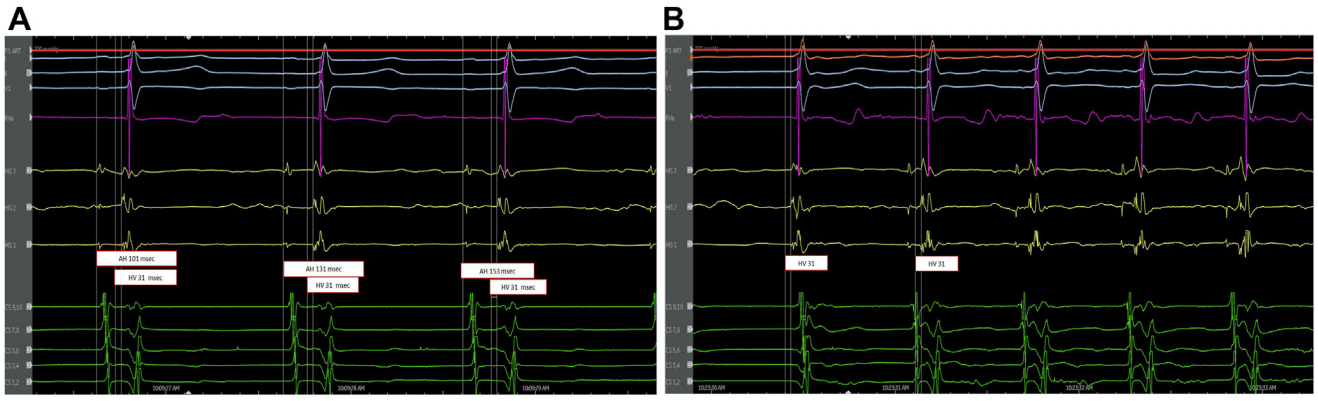

Figure 3 **A:** An incremental increase in AH interval following adenosine, with unchanged AH interval at 31 ms. **B:** A junctional rhythm with an unchanged preexcitation pattern and stable HV interval at 31 ms. AH = atrium-his; CS = coronary sinus; HV = his-ventricle; RV = right ventricle; V = ventricle.

tachycardia.[3] Nonetheless, acquired accessory pathways have been described sporadically over various periods, primarily in patients with congenital heart disease following surgical repairs. A notable example is found in patients with tricuspid atresia who underwent atrio-infundibular connection, known as Fontan surgery, and subsequently developed accessory pathways along the surgical lines between the right atrium and right ventricle.[4,5] Several mechanisms have been proposed, although all remain speculative.

In our patient, we observed dramatic changes in her ECG between the preoperative and postoperative phases, which have been monitored regularly. Her EP study result for risk stratification revealed the presence of an FV pathway without any proarrhythmic properties, and no other inducible SVTs were observed. The FV pathway, conceptually known as a benign entity, is not a true atrioventricular connection; rather, it is a short insertion from the His bundle directly to the myocardium or a deficiency in the usual electrical insulation of the His bundle, bypassing the conventional bundle branches and Purkinje fibers system.

Unlike congenital FV pathways, the pathogenesis of acquired FV pathways is not well established. One hypothesis suggests that the ingrowth of cardiac tissues along surgical lines, particularly in patients with membranous VSD, can disrupt the conduction system, allowing an abnormal extension of His bundle fibers to be directly exposed to the myocardium. This theory is supported by a prior case report where an accessory pathway developed following an orthotopic heart transplant.[6]

Moreover, unavoidable collateral damage resulting from surgery may disrupt the normal conduction systems, potentially revealing previously "concealed" pathways with greater clarity. Not unreasonably, the deduction is supported by Mahmud et al.,[7] who described the enhanced manifest presentation of FV pathway after His pacing. In fact, it is possible that the true prevalence of FV pathway could be undercounted. Furthermore, abnormal healing processes following surgery could alter tissue properties to become more electrically conductive, as described by Peinado et al.[4]

Recently, there was a case series describing acquired FV pathways following repairs of congenital heart disease.[1] Similar to our case report, all patients in the case series underwent VSD repair for conotruncal VSD and their EP study confirmed the presence of FV pathway without any inducible SVT. Given the available data on this type of acquired FV, it is reasonable to deduce that even in its acquired form, FV pathway is benign.

Conclusion

Our case report demonstrated the acquired nature of a preexcitation pattern on ECG subsequent to the repair of a complex congenital heart disease. Subsequently, an EP study confirmed the presence of an FV pathway. Consistent with prior literature, this case report provides additional substantiation for the existence of acquired FV pathways.

Disclosures: The authors have no conflicts of interest to disclose.

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