A case of junctional ectopic tachycardia with demonstration of both HA and HV dissociation during tachycardia

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Introduction

Junctional ectopic tachycardia (JET) is an unusual tachyarrhythmia most commonly encountered in infants and children.[1] These are considered to have origins in the atrioventricular (AV) node or the upper His-Purkinje system (HPS). Langendorf and Pick[2] reported accelerated narrow QRS in the pediatric population that was dissociated from the sinus P wave in 1954 based on surface electrocardiogram (ECG) recordings. However, they attributed these to suppression of the sinus node in conjunction with a mildly accelerated nodal rhythm in the presence of an inflammatory reaction. Langendorf and Mehlman[3] also described premature junctional beats interfering with AV nodal conduction in 1947. The more modern definitions of JET typically involve tachyarrhythmia with rates well over 100 beats per minute. The electrophysiologic (EP) characteristics of JET in the pediatric population were first described by Garson and Gillette[4] in 1979. Multiple subsequent reports have described JET, some of which were treated with ablation.[5–7]

Several EP features have been proposed as suggestive of JET. The most suggestive one is a narrow QRS tachycardia with AV dissociation and a His bundle electrogram preceding each QRS complex.[1,5] On EP testing, the HA intervals during junctional tachycardia tend to be longer than during typical atrioventricular nodal reentrant tachycardia (AVNRT) and may be closer to the HA interval during ventricular pacing.[8] Similarly, an AH-HA response to atrial overdrive pacing is typically observed in JET, as the return beat following cessation of pacing is characterized by an HA sequence.[9] Furthermore, a late-coupled premature atrial complex delivered during His refractoriness does not reset JET, but can alter the subsequent cycle length in AVNRT via the antegrade slow pathway. On the other hand, an early-coupled premature atrial complex capturing the His bundle may advance the tachycardia for 1 cycle in JET, while it often terminates AVNRT.[10] However, the most definitive evidence for a junctional source of such a tachycardia would be the demonstration of both HA and HV dissociation during tachycardia. We report a case of tachycardia presenting with a left bundle branch block (LBBB) morphology, demonstrating HA and HV dissociation in tachycardia during EP study.

KEY TEACHING POINTS

- Junctional ectopic tachycardia (JET) is an uncommon narrow complex tachyarrhythmia most commonly encountered in infants and children.
- Features of JET during electrophysiological study include normal or prolonged HV interval; an AH-HA response to atrial pacing; no change in tachycardia cycle length with a late-coupled premature atrial complex (PAC) during His refractoriness; and possible advancement of the tachycardia for 1 cycle with delivery of an early-coupled PAC.
- The most definitive evidence for a junctional source of tachycardia comes from the demonstration of both HA and HV dissociation during tachycardia.

Case report

A 30-year-old woman presented to our outpatient clinic describing a 3-year history of palpitations. She had undergone an EP study at an outside facility 4 months prior, demonstrating both narrow QRS, LBBB, and right bundle branch block (RBBB) tachycardia, all showing evidence of

KEYWORDS Junctional ectopic tachycardia; Narrow complex tachycardia; VA-dissociated tachycardia; Radiofrequency ablation; Wide complex tachycardia

(Heart Rhythm Case Reports 2021;7:333–337)

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https://doi.org/10.1016/j.hrcr.2021.02.010

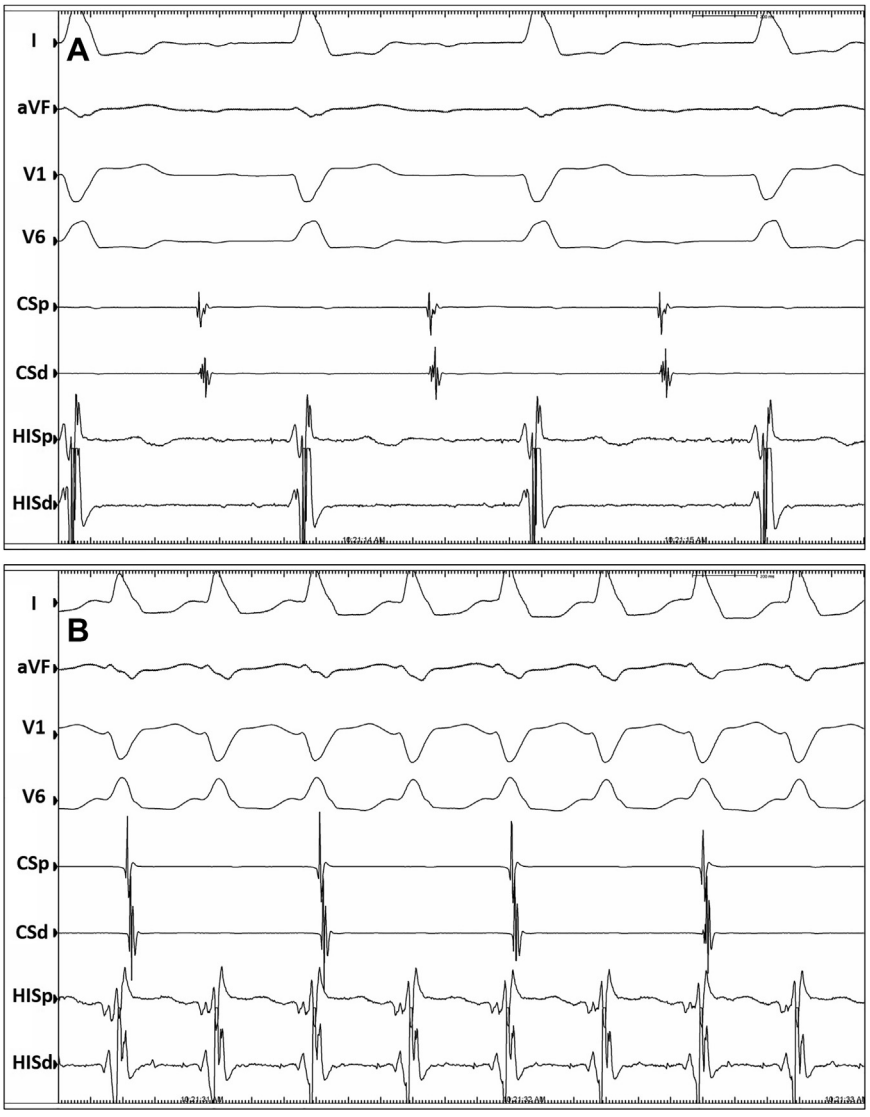

Figure 1 Baseline conduction intervals. **A:** During sinus rhythm. **B:** During tachycardia. Tachycardia was readily and reproducibly induced with burst pacing from the right ventricle. The AH and HV intervals in sinus rhythm were 160 and 51 ms, respectively. HV intervals in tachycardia were unchanged from those in sinus rhythm. The tachycardia cycle length was 300 ms. Note dissociation of His activation during tachycardia from atrial activation. CSp and CSd = proximal and distal coronary sinus electrogram; HISp and HISd = proximal and distal right-sided His/right bundle electrograms.

VA dissociation. An ablation procedure was subsequently performed targeting presystolic fascicular potentials over the left ventricular septum, rendering the tachycardia noninducible. The evening post procedure, she was noted to have new onset of LBBB in sinus rhythm as well as recurrent symptomatic tachycardia with the same LBBB QRS morphology. During the subsequent months she continued to experience frequent recurrences that were suppressed with flecainide. However, wishing to avoid long-term medications, she presented to our institution for another possible ablation.

She presented to the EP laboratory in normal sinus rhythm with LBBB (QRS duration of 125 ms). Under conscious sedation, an EP study was performed using standard pacing and recording techniques. Baseline conduction intervals during sinus rhythm are shown in Figure1A. Tachycardia was readily and reproducibly induced with burst pacing from the right ventricle (RV). Figure 1B shows surface ECGs and intracardiac electrograms of the tachycardia. Episodes of transient complete heart block (CHB) were elicited with catheter manipulation around the right bundle electrogram, likely from mechanical trauma. During episodes of CHB while she remained in sinus rhythm in the atria, repeated right ventricular burst pacing failed to induce the tachycardia. With return of right bundle conduction, the tachycardia again became readily induced. This observation suggests that retrograde conduction into the HPS via the right bundle was essential to initiating the tachycardia. The tachycardia had a cycle length of 300–310 ms, a QRS morphology identical to that in sinus rhythm, and clear VA dissociation (Figure 1B). A His potential preceded each ventricular activation. Entrainment pacing was performed from the RV and left ventricle (LV). Figure 2 shows such entrainment with the associated postpacing intervals (PPI; details in the

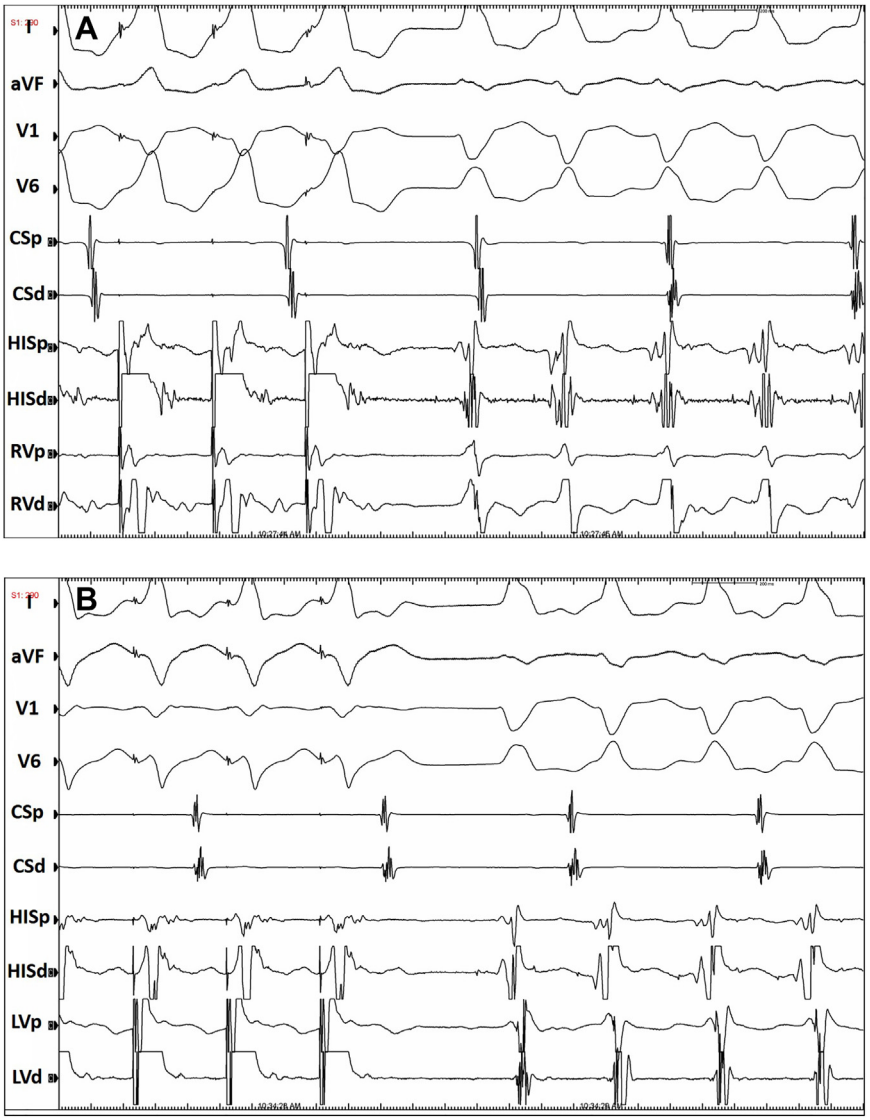

Figure 2 Entrainment during tachycardia from the right ventricle (RV; **A**) and left ventricle (LV; **B**). The tachycardia has a cycle length (CL) of 305 ms. **A:** Entrainment pacing was performed from the distal right-sided His/right bundle electrodes during tachycardia (pacing CL of 290 ms). Local ventricular electrograms showed a postpacing interval (PPI) of 480 ms. HV interval in tachycardia was 53 ms. However, the PPI to the local His electrogram was 400 ms. This shorter PPI would suggest that the His/right bundle was closer to the tachycardia source than the surrounding right ventricular myocardium. **B:** Entrainment pacing was performed from the left ventricular apex. The PPI at the site of pacing was 607 ms, considerably longer than the PPI when pacing from the right. Furthermore, the first tachycardia return beat had earlier His and local RV activation (489 and 549 ms, respectively) than the LV pacing site. These observations are consistent with retrograde left bundle branch block such that pacing from the LV could only penetrate the source of the tachycardia by propagating to the RV and up the right bundle. LVp and LVd were, respectively, proximal and distal electrode pairs on the pacing catheter located near the apical portion of the left ventricle.

figure legend). A mapping catheter was then inserted into the LV via a transaortic approach. Entrainment from the LV apex (Figure 2B) resulted in a longer PPI compared to pacing from the RV (Figure 2A). The left-sided catheter was then withdrawn to a proximal septal position attempting to record a proximal His or left bundle electrogram. At this point, the catheter positioned near the right bundle potential again caused trauma to the right bundle, this time during tachycardia, resulting in HV dissociation while the right- and left-sided His recordings persisted in tachycardia (Figure 3). Figure 3 also demonstrates that the left-sided His recordings activated around 30 ms earlier than the right-sided activation, consistent with the tachycardia source emanating from the proximal left-sided HPS.

A few conservative ablation lesions near the left-sided early His/left bundle deflections were delivered without affecting the tachycardia. However, owing to concerns regarding potential CHB if ablation was pursued, verapamil infusion was tested during tachycardia. During this infusion, tachycardia cycle length prolonged from 300 to 345 ms prior to termination. The tachycardia was no longer inducible at this point. Given the response to verapamil and the concern regarding potential for CHB with further attempts at ablation, it was decided to pursue a treatment

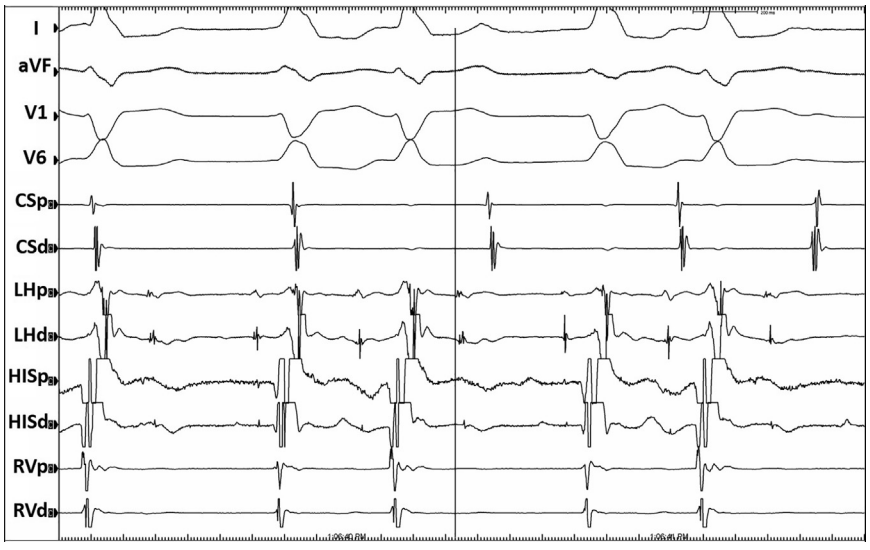

Figure 3 Catheter trauma to the right bundle during tachycardia resulted in dissociation of His activation from ventricular activation. During tachycardia, mechanical trauma to the right bundle resulted in variable HV conduction (3:2 pattern in this tracing) while the right- and left-sided His recordings remained in tachycardia. The left-sided His recording preceded the right-sided one by 30 ms, consistent with the source of the tachycardia being in the left-sided His-Purkinje system. LHp and LHd were, respectively, proximal and distal electrode pairs on the catheter recording from the upper left ventricular septum. The vertical line reveals the timing relationship between the left- and right-sided His activations.

trial with oral medication. Since she responded to flecainide prior to our procedure, it was restarted. Clinical follow-up showed that the tachycardia was still well suppressed on oral flecainide.

Discussion

Junctional tachycardia can emanate from the AV node or from the proximal HPS. In this patient, prior ablation likely generated the LBBB when radiofrequency lesions were delivered in the LV. Prior to ablation, her sinus rhythm 12-lead ECG was normal with a narrow QRS. While ECGs of her tachycardia showed a variety of QRS morphologies prior to any ablation, including narrow QRS, RBBB, and LBBB, her tachycardia following the ablation procedure only showed an LBBB QRS. These observations suggested a single source of tachycardia prior to ablation, likely in the upper portions of the HPS, manifesting various functional bundle branch blocks. Our EP study showed that the left-sided His/ left bundle potential was earlier than the right-sided His/ right bundle potentials during tachycardia, thus further localizing the source to the upper portions of the left bundle branches. Observations during our EP study showed that the LBBB was complete in both directions: (1) catheter trauma inducing RBBB caused CHB; and (2) entrainment pacing during tachycardia from the LV had longer PPI than pacing from the RV, despite the likely source of the tachycardia being from the left-sided proximal HPS. Thus, the source of the tachycardia was from the left-sided proximal HPS, which was only able to propagate to the ventricle via the His and right bundle. The site of the complete LBBB was likely below the left-sided source of the tachycardia. When RBBB occurred in sinus rhythm, causing CHB, the tachycardia became noninducible with ventricular pacing despite its being readily and reproducibly induced prior to the onset of RBBB. This is consistent with the inability of ventricular impulses to penetrate into the source of the tachycardia to initiate it. Based on the ease of inducing the tachycardia with pacing and its response to both verapamil and flecainide, we would propose that the mechanism is most likely microreentry occurring in the upper portions of the left bundle utilizing verapamil-sensitive slowly conducting tissue connecting branches of the upper left-sided HPS. However, we cannot completely exclude a triggered mechanism. It is likely that the initial ablation performed elsewhere targeted the left bundle potential beneath the reentrant site, generating complete LBBB. Thus, they may have been close enough to the reentrant site to temporarily terminate the tachycardia, but the site quickly recovered shortly after the procedure. Unlike previously described fascicular VTs and ventricular nodal or ventricular Hisian reentrant tachycardia,[11] reentry in this case would not depend on myocardial participation in the reentrant circuit.

The demonstration of HA and HV block during tachycardia is the strongest proof of a junctional source of tachycardia, whether it is a reentrant mechanism or a focal automatic source. To our knowledge, there has not been a prior report of a similar demonstration. While this case demonstrates the existence of such an entity, our observations would be difficult to replicate in the EP lab when assessing other potential junctional tachycardia, as block distal to the His bundle bilaterally during tachycardia would be difficult to reproduce in a reversible manner unless 1-sided bundle branch block was already present. This phenomenon was achieved in our case, at least in part, owing to the presence of complete LBBB likely related to the previous ablation procedure. Therefore, transient RBBB from mechanical catheter trauma resulted in HV dissociation in tachycardia with concurrent HA dissociation.

Conclusion

Demonstration of both HA and HV dissociation during tachycardia provides the most definitive evidence for the diagnosis of junctional tachycardia.

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