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SPOTLIGHT

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Tachycardia with cycle length alternans in Ebstein's anomaly

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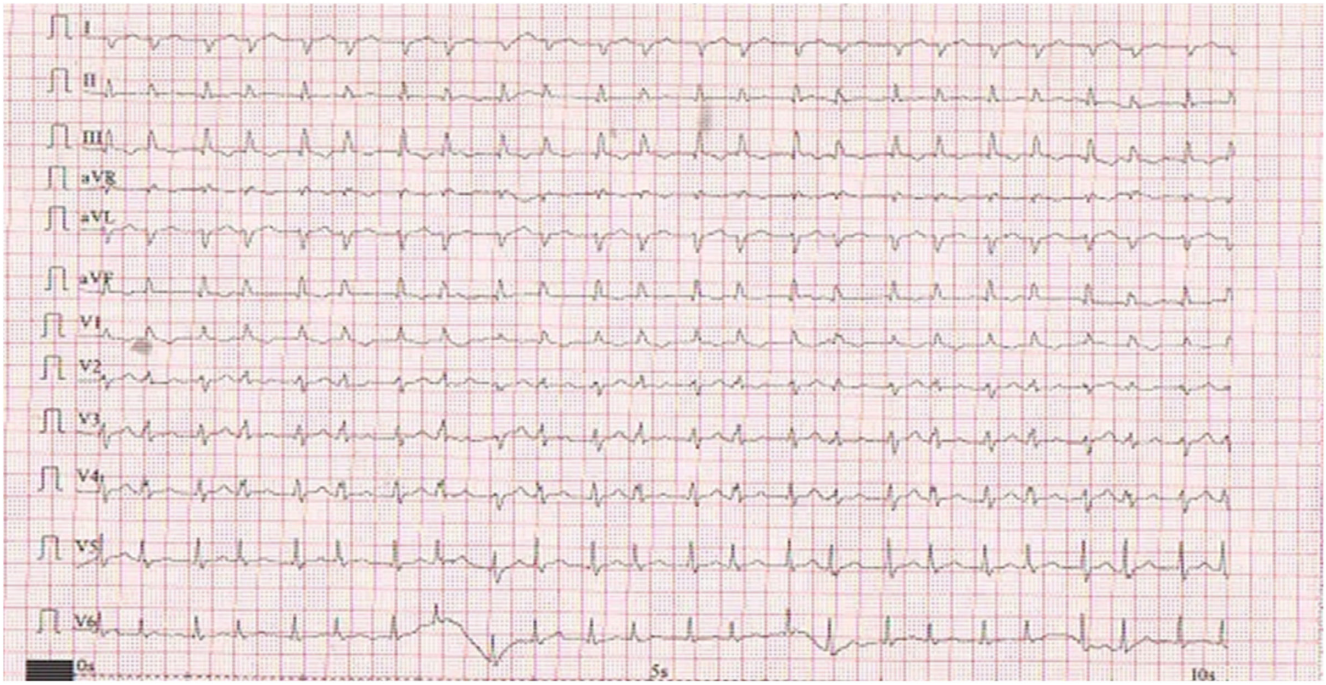
KEYWORDS
congenital heart disease, electrophysiology study, supraventricular tachycardia

Tachycardias in patients with Ebstein's anomaly are commonly AVRT, atrial tachycardia and atrial flutter. A key sign of pre-excitation in a patient with Ebstein's anomaly of the tricuspid valve is the lack of the expected right bundle branch block (RBBB) during sinus rhythm.[1,2]

A 16-year-old girl, known to have Ebstein's anomaly, presented to the arrhythmia clinic with recurrent episodes of paroxysmal palpitations, lasting for several hours and associated with dyspnea. On examination, she had a widely split S1 and a grade 3/6 decrescendo systolic murmur over the left parasternal area. The baseline ECG of the patient showed sinus rhythm with right bundle branch morphology and a PR interval of 130 ms. The ECG of the tachycardia is shown below.

At first glance the tachycardia seems irregular. Closer scrutiny reveals cycle length alternans. The differential diagnoses of cycle length alternans here include (i) atrial tachycardia, (ii) atrial flutter, (iii) orthodromic atrioventricular re-entrant tachycardia (AVRT) using



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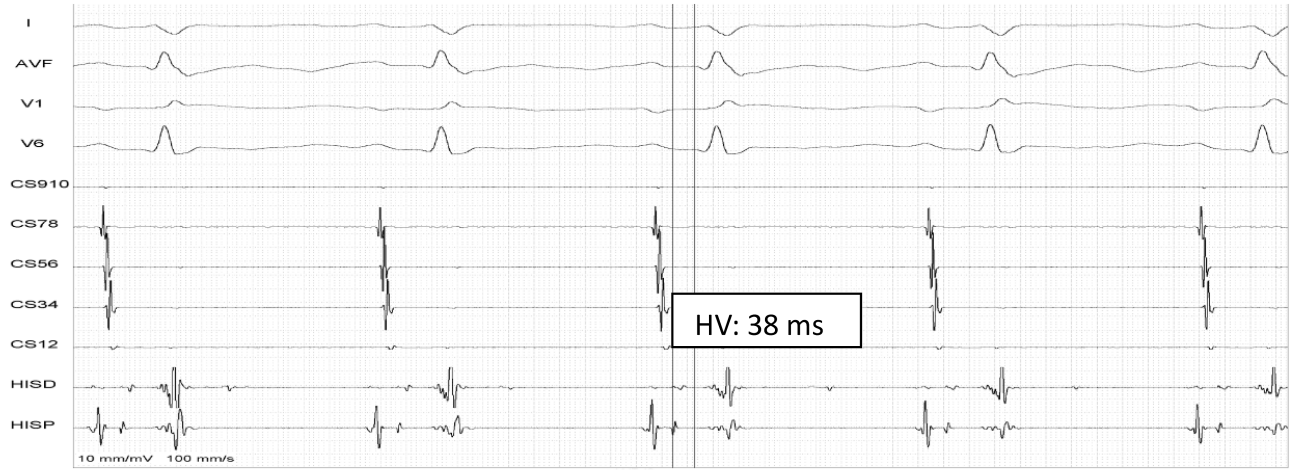

FIGURE 1 Baseline intervals. The figure shows measurement of HV interval at the proximal His which was 38 ms indicating the absence of manifest preexcitation in a patient with Ebstein's anomaly.

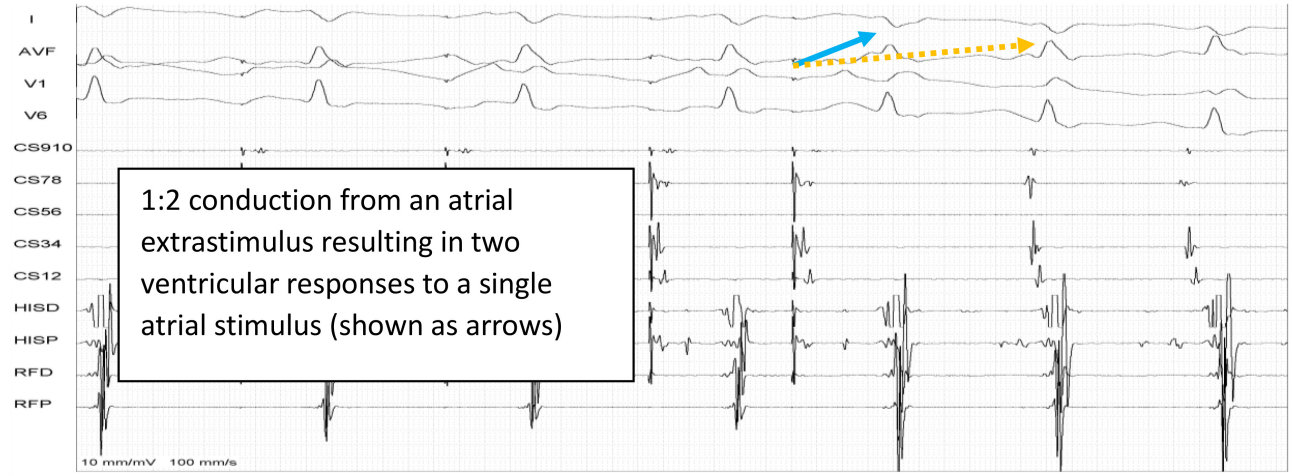

FIGURE 2 Tachycardia induction with A2 showing 1:2 ventricular response to atrial extrastimulus. This was ill-sustained and terminated after three beats because of a premature atrial complex (not shown).

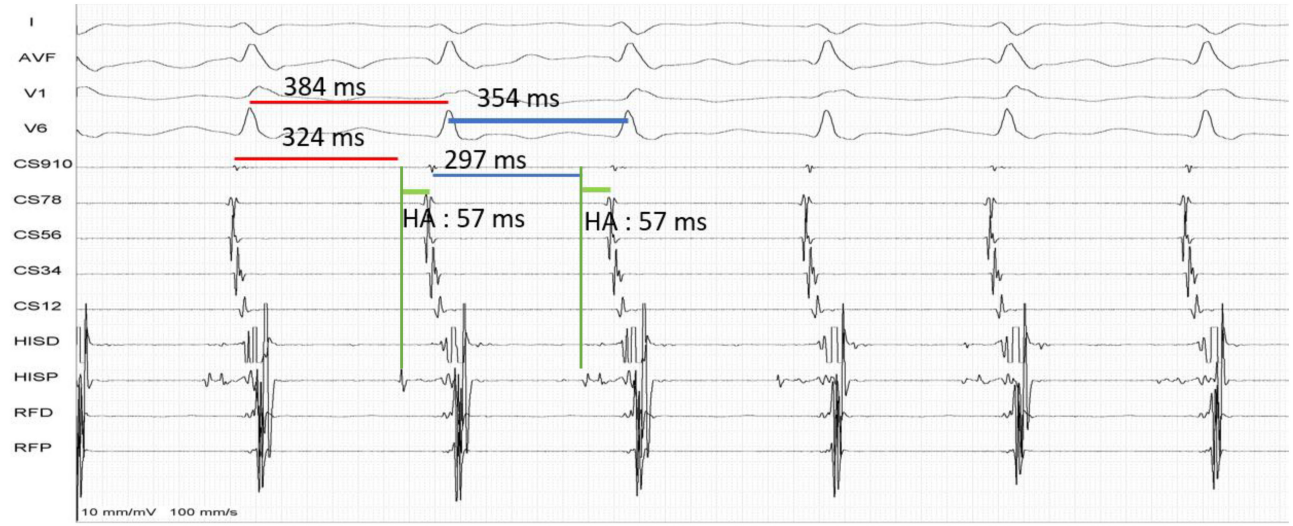

FIGURE 3 Intracardiac electrograms during tachycardia. The tachycardia has a H-A-V sequence with an 'A on V' pattern. The cycle length alternans is because of AH interval alternation while the HA interval (measured here at the proximal CS) remains constant.

two AV nodal pathways, (iv) orthodromic AVRT using two accessory pathways, (v) orthodromic AVRT with longitudinal dissociation of the accessory pathways, (vi) atrioventricular nodal re-entrant tachycardia (AVNRT) with two antegrade slow pathways,[1,2] (vii) AVNRT with two retrogradely conducting pathways and (viii) junctional ectopic tachycardia.[3–5]

An electrophysiology study was performed. Diagnostic quadripolar catheters were inserted into the right ventricular apex and His-bundle region, and a decapolar catheter was inserted into the coronary sinus. The PR interval was 127 ms, the AH interval was 74 ms, the HV interval was 38 ms (Figure 1) and the AV Wenckebach cycle length was 250 ms. The antegrade and retrograde AV nodal refractory periods were both <240 ms. The retrograde conduction was concentric with VA Wenckebach cycle length being 350 ms. No AH, HA or VA jumps were demonstrable.

Tachycardia was induced with a single atrial extrastimulus following a basal train. The extrastimulus displayed a 1:2 conduction, continuing as an 'A on V' tachycardia, with septal VA of −30 ms (Figure 2). The AH interval during the tachycardia was more than 200 ms and the earliest retrograde atrial activation site during the tachycardia was in the His-bundle region indicating retrograde conduction through the fast pathway suggesting the diagnosis of slow fast AVNRT.

The tachycardia showed a cycle length alternans of 384/354 ms. The AH interval alternated between 327 and 297 ms while the HA interval remained constant at 57 ms when measured at the proximal CS (Figure 3). The mechanism of cycle length alternans in AVNRT has been hypothesized to be non-uniform anisotropy because of functional differences in slow pathway circuit, Wenckebach periodicity in AV node or functional lower common pathway, or the presence of multiple AV nodal pathways.

Later, the cycle length became regular at 369 ms. Pacing maneuvers were done during phases of regular tachycardia. A His-refractory ventricular extra-stimulus did not affect the tachycardia. Entrainment from the right ventricular apex ventricle was obtained after 5 captured QRS complexes and was followed by a VAHV response; the corrected VAV-tachycardia cycle length was 132 ms and the stimulus to A-VA was 126 ms. Four rapidly delivered atrial extrastimuli during tachycardia demonstrated AV linking. In view of the above observations, a diagnosis of AV nodal re-entry tachycardia was made. Slow pathway modification was performed. After this, tachycardia could not be induced despite aggressive pacing maneuvers and isoprenaline infusion.

This case highlights the importance of careful analysis of electrocardiograms and electrograms to diagnose the mechanism of tachycardia in patients with Ebstein's anomaly.

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We confirm that we have not received any funding for the above study.

CONFLICT OF INTEREST STATEMENT

Authors declare no conflict of interests for this article.

INFORMED CONSENT

The informed consent from the patient and the legal guardians has been obtained and will be sent should the work be published by the journal.

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