A 23-year-old, otherwise healthy woman presented with recurrent skipped and rapid sustained heart beats. Her physical examination and echocardiogram were normal. The presenting ECG in Figure 1 shows sinus rhythm, a nonconducted sinus p wave, and premature complexes. Both left bundle-branch block (5th and 6th complexes) and right bundle-branch block (10th complex) morphologies are present. The 5th and 10th complexes demonstrate a constant coupling interval and there was no discernable p wave preceding either complex. Although the 5th and 6th complexes appear to be consecutive premature QRS complexes, the timing of the 6th complex is also consistent with an aberrantly conducted sinus beat. This ECG pattern recurred incessantly. The “unexpected” heart block and the premature complexes suggested a diagnosis of His bundle extrasystoles, though this hypothesis did not explain the sustained rapid heart rates. An electrophysiology study was performed.

With recording catheters at the high right atrium, His bundle, and right ventricular apex, the intracardiac correlates of the presenting ECG manifestations were observed. Figure 2 shows an isolated His signal preceding the nonconducted sinus p wave and the previous sinus beat (arrows). This His activation could lead to the atrioventricular (AV) block either by retrograde concealment into the AV node fast pathway or by rendering the His-Purkinje system refractory to the next sinus beat. The apparent isolated His signal is closer to the 3rd sinus beat compared with the 2nd sinus beat, which may explain the AV block after the former and only AH prolongation after the latter. Evidence for concealed retrograde conduction into the AV nodal fast pathway may be inferred from AH interval prolongation of the 2nd sinus beat. In Figure 3, the first sinus beat is conducted with a normal PR interval. The next beat is a wide QRS complex (left bundle-branch block pattern) preceded by a His signal, but without preceding atrial activity. This pattern recurred after the next sinus beat but with 2 consecutive aberrantly conducted QRS complexes. The long HV interval makes a premature ventricular complex unlikely. The working diagnosis was His bundle extrasystoles causing physiological AV block in Figure 2 and anterograde conduction with left bundle-branch aberration in Figure 3. Sinus rhythm with dual His response through fast and slow AV nodal pathways with intermittent aberrant conduction was also considered in the differential diagnosis.

Dual AV node physiology was demonstrated during atrial extrastimulus pacing, and retrograde conduction was absent during ventricular pacing. Isoproterenol infusion (1 μg/min) resulted in dual AV nodal response or junctional complexes initiating a narrow QRS complex tachycardia at a cycle length of 400 ms (Figure 4). The earliest atrial activation occurred at the His position (VA interval, 30 ms), AH interval changes preceded and predicted AA intervals, and ventricular pacing led to a V-A-V response. A premature atrial complex timed to His bundle activation delayed the next His, excluding focal junctional tachycardia and establishing the diagnosis of AV node reentry tachycardia (AVNRT). Cryoablation of AV nodal slow pathway eliminated AVNRT as well as the premature complexes and the apparent His extrasystoles. The diagnosis of dual ventricular response from fast and very slow AV nodal pathways rather than His bundle extrasystole was established, based on the ablation results.
Figure 2. Sinus rhythm with dual His response causing physiological atrioventricular block. Sinus arrhythmia leads to slight acceleration of sinus rates. The first 2 sinus beats conduct to the ventricle with prolongation of AH interval, and an isolated His bundle signal (arrows) is seen after each ventricular activation; the 3rd sinus beat fails to conduct to the ventricle. See text for discussion. Intervals shown in rows are AA, HH, and AH, in milliseconds. HRA indicates high right atrium; HBEP, His bundle proximal; HBED, His bundle distal; and RV, right ventricle.

Discussion

Dual ventricular response to sinus beats due to simultaneous fast and slow AV nodal pathway conduction is a rare phenomenon. This case illustrates the variations in its clinical manifestations and the difficulty in differentiating it from His bundle extrasystoles. His bundle extrasystoles may be concealed, causing physiological heart block, or manifest, typically associated with premature beats. To our knowledge, this is the first reported case of dual ventricular response manifesting as heart block. The diagnosis was ultimately established, based on results of the AV nodal slow pathway ablation. Although the possibility that ablation at a single site in the posterior septum successfully eliminated the AV node slow pathway and the focus of His extrasystoles cannot be definitively excluded, it is unlikely because these are anatomically distinct locations. The electrograms at the slow pathway ablation site in this case were typical with small atrial electrogram, large ventricular electrogram, and no His signal.

His bundle extrasystoles manifest as “unexpected” heart block or PR interval prolongation and premature atrial and ventricular complexes. Intracardiac recordings can confirm the diagnosis by demonstrating spontaneous His bundle depolarization signals. As illustrated in this case, demonstrating that the His signals are indeed spontaneous can be difficult. A seemingly isolated His signal suggesting spontaneous His depolarization was present in this case (Figure 2) but was due to dual AV nodal conduction with block below the His. Block below the His can be explained by the preceding variability in H-H intervals causing a long-short sequence (Figure 2).

A shorter HV interval during premature beats compared with sinus beats and a distal to proximal activation sequence of His electrograms establishes His bundle origin of the beat. However, these findings may be present only in cases of extrasystole originating from the distal His or bundle branches. Other observations could be suggestive but not diagnostic of either condition. Absence of a demonstrable dual AV node physiology during electrophysiology study should favor His bundle extrasystole. Consistent reproduction of the rhythm by atrial stimulation may suggest dual AV nodal response. The coupling interval of the premature beat can vary in both conditions. Patients with dual AV nodal response should have unidirectional retrograde conduction block in the slow pathway to prevent retrograde invasion from fast pathway conduction.2 This can be inferred if ventricular pacing with retrograde fast pathway conduction at slow cycle lengths induces AV nodal echo beats or AVNRT.

This case report illustrates the difficulty in differentiating sinus rhythm with dual AV nodal ventricular response from His bundle extrasystoles. Clinicians may be faced with a scenario in which the differentiation of dual AV nodal response and His bundle extrasystole cannot be established with certainty and may have to rely on suggestive features of either diagnosis. In our patient, the diagnosis was confirmed only after elimination of all the manifested arrhythmias with AV nodal slow pathway ablation. Dual AV nodal ventricular response, a condition readily amenable to ablation, should be routinely considered in the differential diagnosis of His bundle extrasystole.

Disclosures

None.

References


Key Words: dual response • His bundle extrasystole • heart block • ablation • electrophysiology
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