

Atypical accessory fibers as a lone or additional substrate for 1:2 response phenomenon?



Evgenii Labartkava, MD, Giorgi Chkholaria, MD, Aleksandre Chkholaria, MD

From the Cardiology Department, Kutaisi Central Hospital, Kutaisi, Georgia.

Introduction

The phenomenon of a “double fire,” double atrioventricular nodal (AVN) response or dual AVN nonreentrant tachycardia (DAVNNT) first described in 1975 and published until the present mainly as isolated case reports, is a rare clinically manifested arrhythmia. Specific electrocardiographic sign of this infrequent phenomenon is that 1 sinus beat is followed by 2 narrow or wide (as a result of aberrancy) QRS complexes^{1,2} owing to the simultaneous anterograde conduction via fast and slow pathways (FP, SP) of the functionally dissociated AVN. Atypical bypass fibers with decremental conduction is another rare clinical entity. Electrocardiographic recognition of those structures during sinus rhythm (SR) may be difficult owing to the presence of subtle or no preexcitation. Premature ventricular complexes (PVC) ablated from Koch’s triangle is also an infrequent entity.³ Crossing key points for all of them might be QRS linking, atypical accessory fibers, or crux cordis, making differential diagnosis challenging. We describe a case of idiopathic frequent monomorphic PVCs from the basal ventricular septum with a possible diagnosis as 1:2 response phenomenon over atypical accessory fibers.

Case report

A 60-year-old man with a history of severe complaints for the past 2 years was diagnosed with intermittent arrhythmia as frequent PVC. He was recommended treatment with antiarrhythmic drugs, which was ineffective, and the patient referred to our hospital for further evaluation.

The patient experienced frequent episodes of palpitation and chronic cough on admission. Echocardiography demonstrated good ventricular function, and baseline electrocardiogram (ECG) showed SR without preexcitation and frequent wide-QRS premature beats (PB) with left bundle branch block morphology and superior axis, initially verified as PVCs. The 24-hour Holter monitoring revealed frequent monomorphic wide-QRS PBs, alternating repetitive episodes

of bigeminy and trigeminy, complete absence of any PBs during sleep, and PB burden of 29%. During arrhythmogenic activity (present only when preceding SR was in a range of 71–84 beats/min), P-wave to QRS ratio were observed in the following settings: most frequently alternating 1:1, 1:2, and 1:0 sequence; less frequently alternating 1:2 and 1:0 sequence. During careful analysis of Holter tracings we revealed constant and fixed coupling intervals (CI) of PBs to previous QRS complex ($\Delta CI = 0$ ms) and regular irregularity (Figure 1, Supplemental Figures S1–S3). Depending on the 12-lead PB QRS morphology, we predicted the site of ventricular exit from the subtricuspid septum of the right ventricle (RV). Based on this we hypothesized 3 possible diagnoses: PVCs, automaticity from bypass fibers, or 1:2 AVN response with bystander accessory fibers. After written informed consent was obtained, electrophysiological study (EPS) and radiofrequency ablation (RFA) were performed in the postabsorptive, unsedated antiarrhythmic drug-free state.

Three diagnostic electrodes and a mapping catheter were inserted and positioned in the following sites: high right atrium, coronary sinus, His bundle region, and subtricuspid septum of the RV. Atrio-His and His-ventricle intervals were within the normal range (80 ms and 44 ms, respectively) during SR. We experienced complete disappearance of any arrhythmogenic activity during attempts to provide anterograde/retrograde programmed stimulation (decremental or extrastimulus testing,) from any sites (coronary sinus/right atrium and RV base / RV apex) at multiple S₁-S₁ cycle lengths. Anterograde conduction over FP+AVN+His-Purkinje system with gradual increase in A-H interval without manifestation of discontinuous curve, double AVN physiology, or induction of any tachycardias were registered during all times. Anterograde effective refractory period (AERP) and anterograde Wenckebach of the AVN were within the normal range (280 ms and 320 ms, respectively). Retrograde stimulation revealed V-A dissociation. Intravenous infusion of isoproterenol did not reveal any change. The activation mapping confirmed ventricular exit site of PBs in the posteroseptal para-Hisian space of the RV at the subtricuspid level. Subsequently we made careful mapping of Koch’s triangle and the surrounding region. A discrete low-frequency electrogram preceding each PB (later verified as M-potential) was recorded at the posteroseptum at the

KEYWORDS 1:2 AVN conduction; Mahaim fibers; Pre-excitation; Premature ventricular complexes; Crux cordis; Radiofrequency ablation (Heart Rhythm Case Reports 2020;6:733–737)

Conflict of interest: All authors declare that there is no conflict of interest.
Address reprint requests and correspondence: Dr Evgenii Labartkava, Cardiology Department, Kutaisi Central Hospital, Solomon Pirveli 10, Kutaisi 4600, Georgia. E-mail address: Eugene-geo@mail.ru.

KEY TEACHING POINTS

- “Double fire” might be a heterogeneous group of arrhythmias with different substrates, mechanisms, and electrocardiogram (ECG) manifestation in which 1:2 atrioventricular nodal (AVN) response composes only some part. Anatomical and physiological substrates for 1:2 response phenomenon are functional dissociation of the AVN (with specific conditions) as a solo entity, accessory Mahaim fibers as another distinct entity, or a combination of both. This may explain the presence of 1:2 response phenomenon in similar proportion of male and female patients compared to AVN reentrant tachycardia.
- In our case it was impossible to manifest anterograde conduction over bypass structures during atrial pacing, but the accurate preliminary analysis of electrocardiogram (ECG) allowed us to suspect Mahaim fibers and carry out mapping of Koch’s triangle. As a consequence, registered M-potential, along with the results of pacing (selective capture) and radiofrequency ablation, confirm the diagnosis. Based on this we made the following conclusion: while it is impossible, with atrial pacing maneuvers, to validate conduction down the Mahaim fibers during the electrophysiology study, this fact does not exclude the origin of atypical accessory fibers above the AV ring.
- ECG and electrophysiological signs of 1:2 response over Mahaim fibers resembles premature ventricular contraction with exit site at the para-Hisian space. High burden and ineffective antiarrhythmic drug treatment promote ablation therapy as a treatment of choice. The described specific properties and manifestations should allow one to suspect atypical accessory fiber as a substrate in similar cases and carry out correct planning of cardiac mapping procedures, avoiding “unnecessary” high-risk application at the para-Hisian space.

hinge of the septal leaflet of the tricuspid valve (Figure 2). This M-potential recording site was far enough from the site of registration of the most proximal His bundle electrogram (more than 10 mm) and revealed the following constant intervals: fixed $A_{\text{His}}\text{-M}$ interval = 590 ms (atrial electrogram from His bundle electrode to M-potential interval), fixed H-M interval = 500 ms (His electrogram to M-potential interval), fixed M-V interval = -45 ms (M-potential interval to QRS_{V1} interval) (Figure 2).

Based on the manifested parameters, we further declined toward the presence of atypical accessory fibers with decremental conduction—Mahaim fibers. There were 2 main questions to address: (1) the proximal insertional site of Mahaim fibers as nodal or atrial; and (2) the origin of wide-QRS PB as fully preexcited or fusion beat as a result of collision of 2 wavefronts—1 over Mahaim fibers and the second over the His-Purkinje system.

Our attempts to manifest preexcitation during pacing from different sites of Koch’s triangle did not achieve success—during all times we had capture of atrial myocardium with subsequent conduction over FP. When pacing from the site of registration of M-potential, we registered simultaneous capture of the atrial myocardium and possible accessory fibers followed by “pre-existing” QRS complexes and stimulus-to-QRS interval in lead V1 ($\text{St.-QRS}_{\text{V1}}$) equal to M-potential to QRS_{V1} interval (Figure 3).

It was impossible to register any His or proximal right bundle branch potential subsequently preceding or following wide-QRS complexes (absent or inscribed within local ventricular electrograms) and thus to differentiate between anterograde and retrograde capture of the His-Purkinje system. The attempts of RFA at ventricular exit site were noneffective.

One-point ablation at the M-potential registration site was effective. After 5 seconds of starting RFA we registered disappearance of any PBs; at 10 seconds of RFA we registered a single junctional beat with V-A dissociation. During the full RFA time—60 seconds with maximum temperature 55°C and power 45 W—we registered a total of 3 junctional beats, all with V-A dissociation (Supplemental Figure S4). Results of RFA further support our diagnosis, but do not allow full exclusion of PVCs.

After RFA during anterograde and retrograde stimulation, the registered conductive properties did not differ from the ones evaluated before ablation.

Follow-up electrocardiogram tracings and Holter monitoring at 3, 6, and 9 months did not reveal any arrhythmogenic activity. The patient remains free of symptoms and complaints.

Discussion

QRS linking (fixed CI of PBs with $\Delta\text{CI} = 0$ ms) suspects the mechanisms of arrhythmogenesis as an echo beat (reentrant) or double fire. We noticed the similarity of the above-mentioned patient with the cases of PBs verified as PVCs from the basal septum and successfully ablated from Koch’s triangle.³ Differential diagnosis between them is challenging. But as described above, the type of regular-irregularity with the exact coupling of the PBs to the preceding SR ($\Delta\text{CI} = 0$ ms) with a narrow zone of expression could be explained only by the participation of the nodal-like structure with fine autonomic innervation in an arrhythmogenic mechanism. Based on this, along with well-known gold-standard criteria defined to suspect and confirm the presence of double fire, we conclude the diagnosis as 1:2 response over nodoventricular (NV)

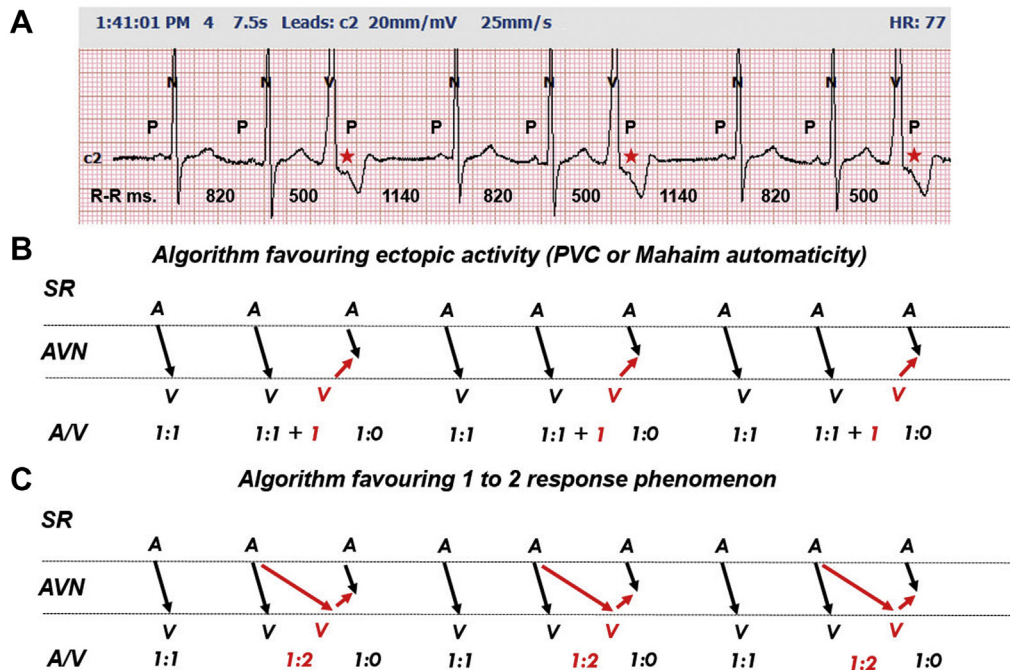


Figure 1 The rhythm strip on channel c2 of Holter monitoring during episodes of trigeminy (A) and ladder diagrams (B and C) demonstrating the proposed mechanisms. **A:** Two sinus beats followed by wide-QRS premature beats (PBs) with fixed coupling intervals (CI = 500 ms) and fixed compensatory pauses (1140 ms). Every third blocked P wave is visible at the end of PBs (red asterisk); P-P intervals are regular and equal to 820 ms. **B:** Ladder diagram demonstrating atrial-to-ventricular ratio (A/V) as 1:1, 1:1+1, 1:0 during rhythm strip on panel A and advising possible diagnosis as premature ventricular contraction or Mahaim automaticity. **C:** Ladder diagram demonstrating A/V as 1:1, 1:2, 1:0 during rhythm strip on panel A and offering possible diagnosis as 1:2 response. Note that, along with fixed CI of PBs to the previous QRS complex (QRS₁-QRS₂), we also have fixed intervals between P waves and QRS₂. So 1 P wave is followed by 2, 1 narrow and second wide-QRS complexes. AVN = atrioventricular node; SR = sinus rate.

fibers, and we present the discussions with the resultant conclusions as follows.

The 12-lead surface ECG is a “gold standard” to suspect an unusual double fire with the strongest significant hint as a single P wave followed by 2 narrow or wide QRS complexes, fixed coupling intervals of QRS extra beats to the previous QRS complex, and so-called regular-irregularity observed during long tracings.¹ Manifest Mahaim fibers dependent on the distal insertion site presented variable QRS expression with no or minimal pre-excitation. Therefore to suspect 1:2 response with concomitant bypass accessory fibers is too difficult to accomplish, and even so, it needs further conclusion during EPS.

The electrophysiological conditions required for 1:2 ventricular response to occur (summarized and published in detail²) are expressed and maintained within a specific range of underlying heart rate and autonomic tonus. For this reason, in the case of the above-mentioned patient 1:2 response was only registered within the specific range of underlying heart rate and completely disappeared during sleep and programmed stimulation.

Manifestation of AVN functional dissociation during programmed stimulation required a specific ratio of functional properties (AERP and Wenckebach) of the fast and slow AVN pathways. Inability to reach (narrow zone) necessary functional differences between these 2 during EPS (AERP_{FP} < AERP_{SP}) explains the failure to unmask the discontinuous curve of AVN anterograde conduction.

In our case it was impossible to fully differentiate whether substrate was atrioventricular (AV) or NV. We are more inclined toward the presence of the last variant out of these 2, based on the location of effective ablation site and registration of junctional beats during heating.

We undertook a few attempts to differentiate between the origin of wide QRS complexes as fully pre-excited or fusion beat as a result of collision of 2 wavefronts: registering and timing any His or proximal right bundle branch potential subsequently preceding or following PB, manifesting discontinuous curve of the AVN before and after RFA, and attempts of initial ablation at the ventricular exit site for blocking of conduction over bypass fibers with preserving conduction over AVN slow pathways. All of them failed. We are more inclined toward a theory of wide QRS complexes being fully pre-excited, since, along with nonrevealed changes in QRS morphology or width of all PBs, only a few single junctional complexes registered during RFA instead of accelerated junctional rhythms, rather insufficient for complete modification of AVN anterograde SP conduction and absence of discontinuous curve of the AVN conduction before and after RFA. (The substrate and proposed mechanism of 1:2 response over NV fibers are illustrated and described in [Supplemental Figure S5](#)).

In the case of the above-mentioned patient, P-wave to QRS correlation as described was in 2 different settings (most frequently 1:1, 1:2, 1:0 sequences). We thought that the site of possible block was located elsewhere in the

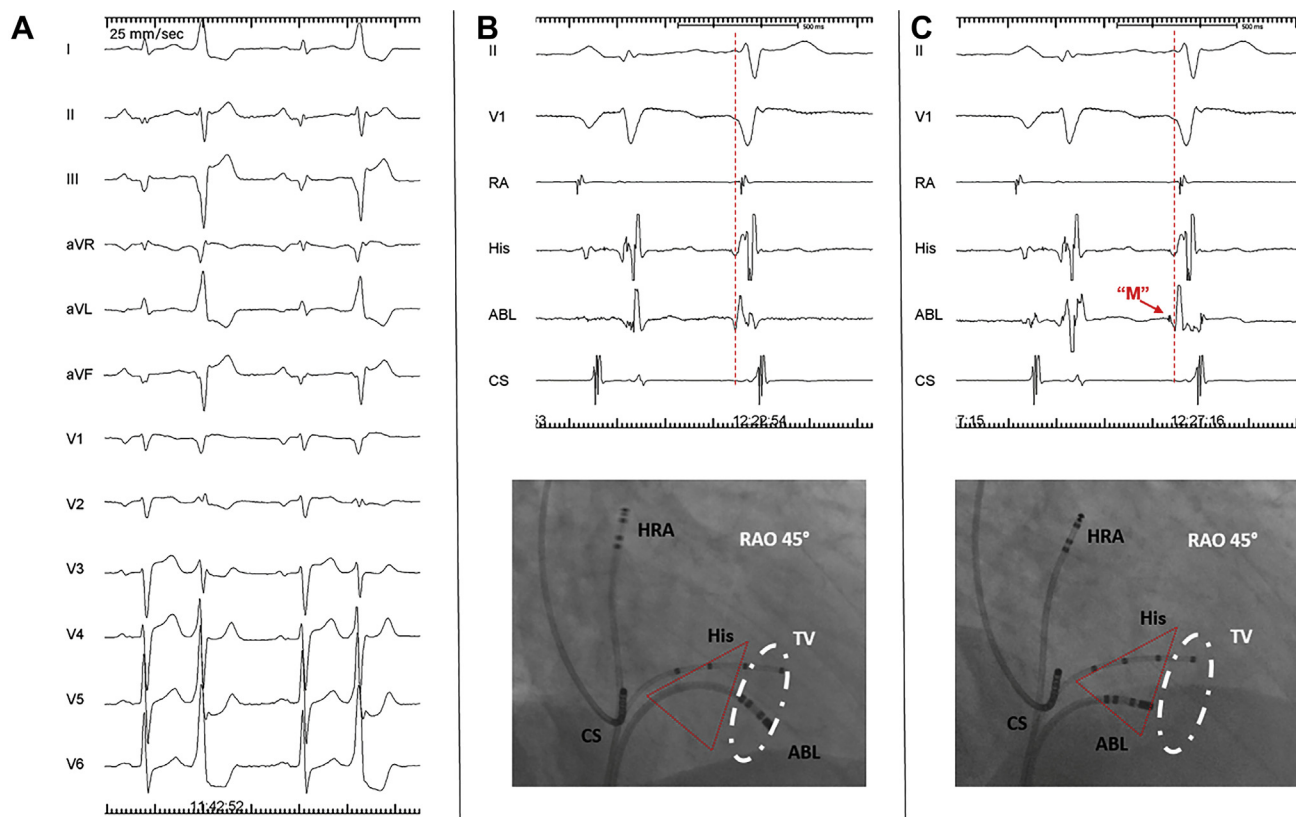


Figure 2 A: Twelve-lead electrocardiogram (ECG) during sinus rhythm and premature beats. B,C: Surface ECG with intracardiac tracing and correspondent radiograph of electrode position within the heart registering ventricular exit site (B) and M-potential (C). Note that local ventricular electrogram on channel ABL at exit site and M-potential precedes the QRS onset by 30 and 45 ms, respectively. Vertical dotted lines indicate the onset of QRS in lead V₁. Red dotted triangle indicates Koch's triangle. White dotted circle indicates tricuspid valve. ABL = mapping catheter; CS = coronary sinus; His = His electrode; HRA = high right atrium; TV = tricuspid valve.

His-Purkinje system, the distal common pathway, or the FP of the AVN as a consequence of concealed conduction.

One sinus nodal origin P wave followed by 2 narrow or wide (aberrant or pre-existing) QRS ventricular responses with fixed intervals P-QRS₁ and P-QRS₂ and revealing regular-irregularity should be a specific, taking into consideration, ECG signs to suspect double fire response phenomenon. Anatomical and physiological substrates for 1:2 response phenomena are functional dissociation of the AVN (with specific conditions²) as a solo entity, accessory Mahaim fibers as another distinct entity, or a combination of both. This may explain the presence of 1:2 response phenomenon in similar proportion of male and female patients¹ compared to AVN re-entrant tachycardia (more frequent in female patients). In the case of AV fiber as a possible substrate for 1:2 response phenomenon, ECG might have a specific manifestation in a fashion of regular-irregularity: 1 sinus P wave contributes to 2 QRS complexes, out of which the first QRS is narrow or wide (aberrancy) and the second one is always wide—pre-excited.

In the case of NV fiber as a substrate for 1:2 response phenomenon, if these fibers possess anterograde conductive properties ECG manifestations might resemble the ones described above during AV fibers. If these fibers are concealed (described until now in only 3 cases as possible

substrate for 1:2 response, verified initially as junctional ectopy or even AVN block^{4,5}), ECG manifestations might resemble all possibilities described during DAVNNT (Supplemental Figure S6).

The accurate preliminary analysis of ECG allowed to suspect double fire over Mahaim fibers and carry out mapping of Koch's triangle. As a consequence, registered discrete potential, along with the results of pacing (selective capture) and RFA, confirmed the diagnosis.

Conclusion

The discovery of this case allows us to make the following conclusions:

- “Double fire” might be a heterogenous group of arrhythmias with different substrates, mechanisms, and ECG manifestation in which 1:2 AVN response composes only some part.
- Registered QRS complexes during “double fire” are possible to be narrow or wide. Wide QRS along with aberrancy is possible to be fully pre-excited or fused complexes. While it is impossible with atrial pacing maneuvers to validate conduction down the Mahaim fibers during the EPS, this fact does not exclude the origin of atypical accessory fibers above the AV ring.

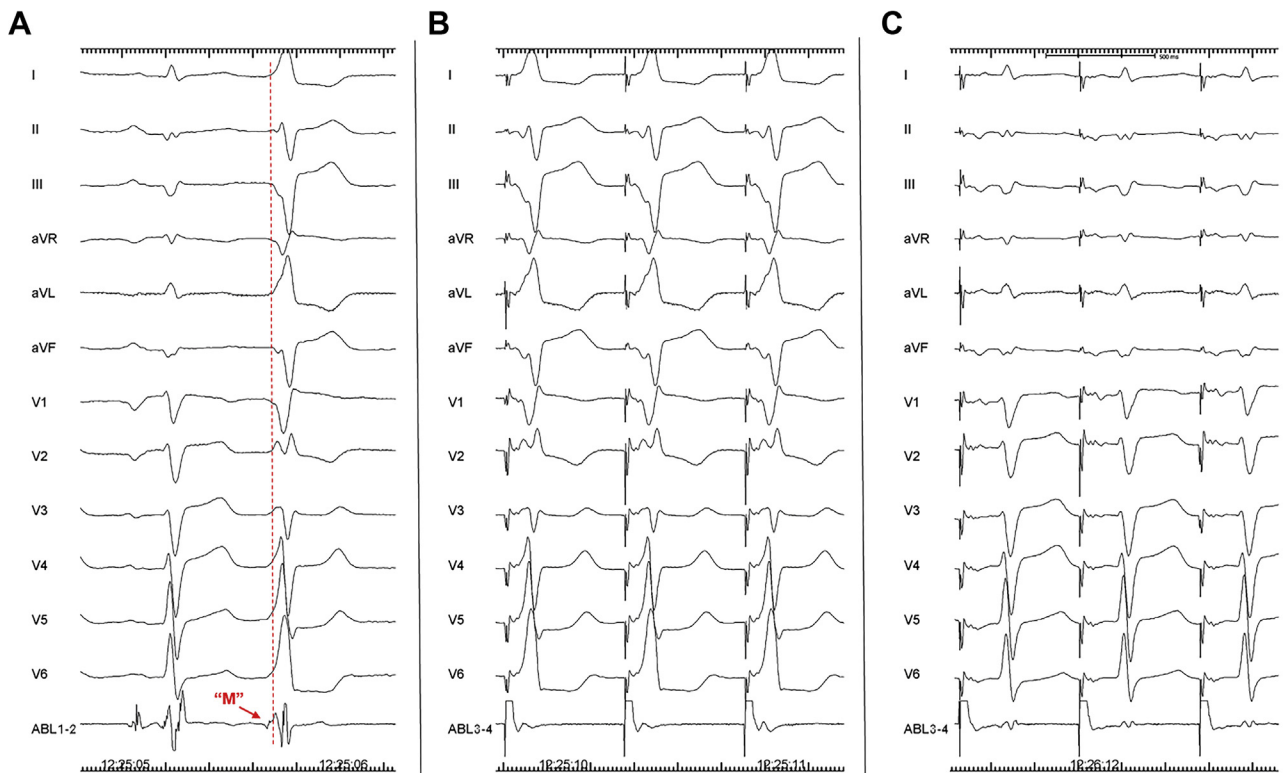


Figure 3 **A:** Twelve-lead electrocardiogram (ECG) during sinus rhythm and wide-QRS extra beats with intracardiac tracing from mapping electrode positioned at M-potential registration site. Vertical dotted line indicate the onset of QRS in lead V₁. M-potential precedes the QRS onset by 45 ms. **B:** Twelve-lead ECG during pacing from mapping electrode positioned at M-potential registration site. Note: Morphology of QRS complexes during pacing resembles the ones described during spontaneous premature beats and St.–QRS_{V₁} interval equal to M-potential to QRS_{V₁} interval. **C:** Twelve-lead ECG during pacing from mapping electrode positioned at posteroseptal region of Koch's triangle. Note conduction over fast atrioventricular nodal pathways with complete disappearance of any arrhythmogenic activity. ABL = mapping catheter; "M" = M-potential; St.–QRS_{V₁} interval = interval between stimulus and QRS in lead V₁.

- The registration of single (or even accelerated rhythm) junctional complexes with V-A dissociation may be a characteristic electrophysiological sign of RFA during 1:2 response.

Appendix Supplementary data

Supplementary data associated with this article can be found in the online version at <https://doi.org/10.1016/j.hrcr.2020.07.004>.

References

1. Peiker C, Pott C, Eckardt L, et al. Dual atrioventricular nodal non-re-entrant tachycardia. *Europace* 2016;18:332–339.
2. Wang N. Dual atrioventricular nodal non re-entrant tachycardia: a systematic review. *Pacing Clin Electrophysiol* 2011;34:1671–1681.
3. Santangeli P, Hutchinson MD, Supple GE, et al. Right atrial approach for ablation of ventricular arrhythmias arising from the left posterior–superior process of the left ventricle. *Circ Arrhythm Electrophysiol* 2016;9:e004048.
4. Tuohy S, Saliba W, Pai M, Tchou P. Catheter ablation as a treatment for atrioventricular block. *Heart Rhythm* 2018;15:90–96.
5. Chung R, Wazni O, Dresing T, et al. Clinical presentation of ventricular-Hisian and ventricular-nodal accessory pathways. *Heart Rhythm* 2019;16:369–377.