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CASE REPORT

Successful ablation of atrioventricular nodal reentry tachycardia following the atriopulmonary Fontan procedure

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Arrhythmia is a prevalent complication of the atriopulmonary Fontan procedure, most commonly macro-reentrant tachycardia within the right atrium as a consequence of long-term morphological and haemodynamic changes coupled with surgical intervention. We describe a patient with a long history of supraventricular arrhythmia following the atriopulmonary Fontan procedure identified as atrioventricular nodal reentry tachycardia at electrophysiological assessment. Despite anatomical and technical difficulties related to right atrial morphological changes and loss of the tricuspid annulus with its associated anatomical landmarks, the arrhythmia was successfully ablated using a combined antegrade and retrograde mapping approach.

Case report

An 18-year-old man who had undergone a one-stage atriopulmonary Fontan procedure aged 3 years for classical tricuspid atresia, presented with sustained tachycardia. His first episode of atrial arrhythmia was documented at the age of 7, from which time he had been treated with sotalol. Until recently, he reported a good quality of life although had suffered lately with both lethargy and exercise intolerance, but no previous specific symptoms of palpitation. The 12-lead ECG demonstrated a regular, narrow complex tachycardia with a ventricular rate of 140 bpm (*Figure 1*). The atrioventricular relationship was 1:1 with the P wave seen immediately following the QRS complex. The arrhythmia persisted despite DC cardioversion and amiodarone therapy and he was transferred to our institution for electrophysiological (EP) assessment.

In view of the likelihood of intra-atrial reentry as the mechanism, we elected to use electroanatomic mapping (CARTO/Biosense Webster, Diamond Bar, CA, USA) during the EP study. Earliest atrial activation during tachycardia was immediately superior to the His bundle, spreading initially superiorly, and subsequently inferiorly along the atrial septum (*Figure 2A*). Two separate wave fronts subsequently progressed medially and laterally around the right atrium, finally colliding on the posterior aspect. The coronary sinus (CS) was activated proximal to distal. Considering the potential diagnoses at this stage, conventional EP assessment was performed. Sinus intervals were of cycle length 1050 ms, AH 88 ms, HV 48 ms. During programmed atrial stimulation at a drive cycle of 600 ms, with single extra stimuli (S2), a decrease in the coupling interval from 420 to 400 ms lengthened the AH interval by 52 ms and was associated with a single atrial echo beat. The clinical arrhythmia could be reliably induced with two atrial extra stimuli (S2 470 ms S3 430 ms) and during tachycardia, the intervals were AH 358 ms, HV 54 ms, and HA 120 ms. Attempts to introduce a His synchronous ventricular premature beat (HSVPB) from the lateral cardiac vein via the CS during tachycardia were unsuccessful. Tachycardia always terminated with antegrade atrioventricular node (AVN) block, either spontaneously or with adenosine. A presumptive diagnosis of typical (slow–fast) atrioventricular nodal reentry tachycardia (AVNRT) was made based on the above findings. Because of the uncertain risk of AVN block, radiofrequency ablation was not attempted and based on the diagnosis attained at EP study, a trial of verapamil was attempted.

Despite initial improvement, the patient suffered from worsening symptomatic arrhythmia and exercise intolerance, with asymptomatic arrhythmia documented on 7-day Holter (Lifecard CF, Delmar Reynolds, Hertford, UK), so a further EP study to define the mechanism was undertaken

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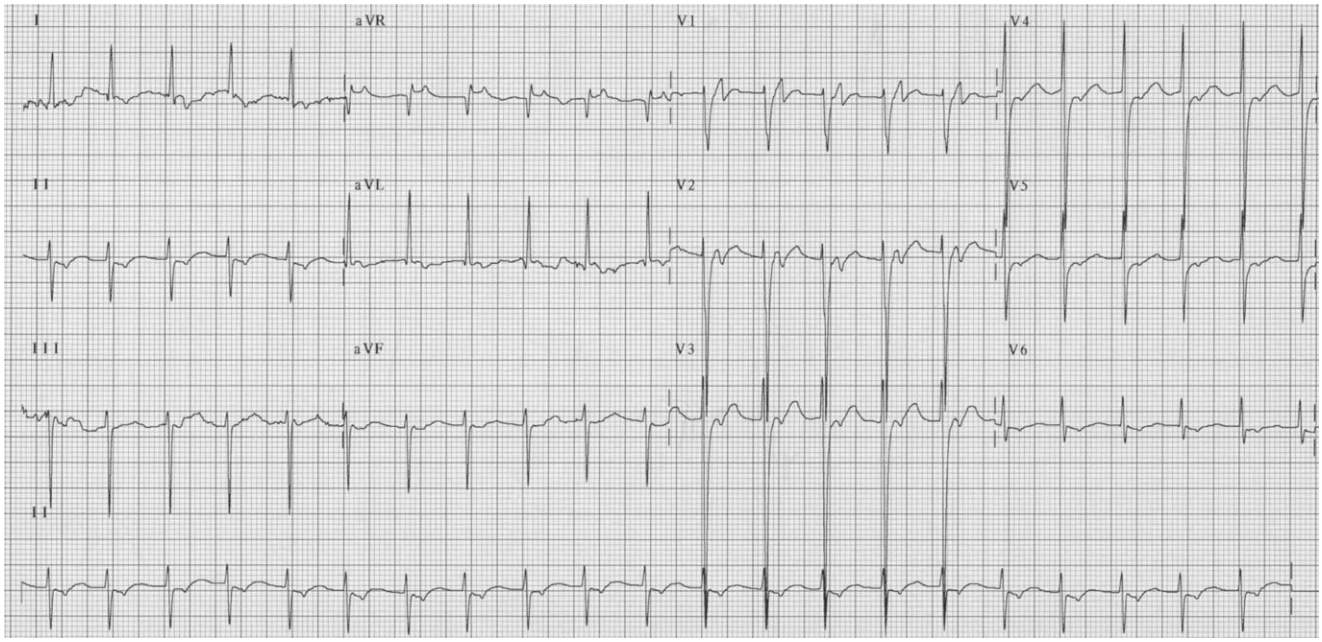

Figure 1 The 12-lead ECG during clinical tachycardia. The atrioventricular relationship is 1:1 with P waves seen immediately following the QRS complex.

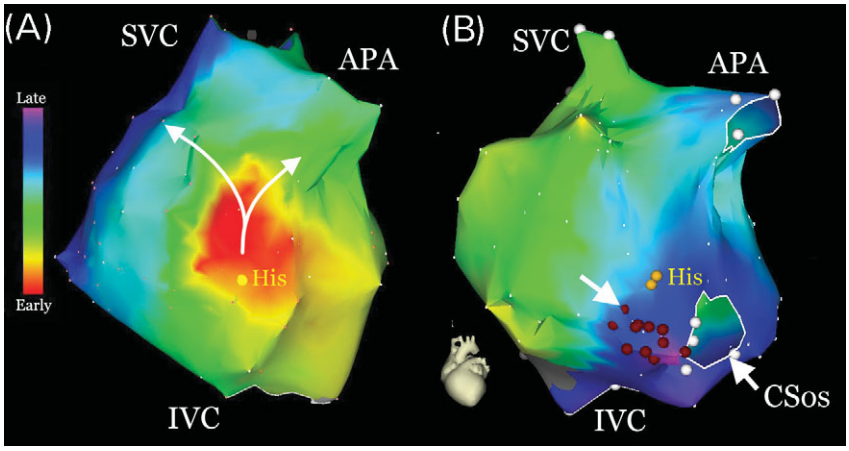

Figure 2 Electroanatomic recreation of the right atrium seen from the left anterior oblique view recorded during (*A*) tachycardia and (*B*) sinus rhythm. Activation is encoded by colours and depicted in the bar on the left-hand side of the image; red is earliest and purple latest activation. The superior caval vein (SVC), IVC, CSos, and atriopulmonary anastamosis (APA) have been annotated. The position of the His bundle electrogram (His, yellow dots) has been marked anterior and superior to the CSos. During tachycardia (*A*) earliest activation can be seen immediately superior to the His bundle in the presumed position of the fast pathway, and progresses initially along superiorly (curved white arrows) and then inferiorly along the atrial septum, following which wave fronts progress both medially and laterally around the chamber ultimately colliding on the posterior wall (not seen). Radiofrequency energy (red dots) was applied during sinus rhythm (*B*) initially adjacent to the CSos and moved superiorly until an accelerated junctional response was seen. The successful lesion is marked with a white arrow.

with a view to radiofrequency ablation. Given the high probability that ablation would be performed close to the position of the His bundle and the difficulty in ensuring His catheter stability in the absence of a tricuspid valve, we elected to use electroanatomic mapping to define this area in the right atrium. A bipolar active fixation reference catheter and quadripolar F-curve mapping catheter were deployed in the right atrium and electroanatomic mapping performed during sinus rhythm. Two further D-curve quadripolar mapping catheters were introduced retrogradely to the left heart, one to record and mark the position of the His signal during mapping and ablation, and the second in the left ventricle for diagnostic assessment and pacing in the event of AVN block. The same tachycardia (cycle length 440 ms) was induced with programmed atrial stimulation (S1 500/S2 360; A1H1 228 ms; A2H2 362 ms) (*Figure 3A*). Programmed ventricular pacing demonstrated decremental conduction via the AVN. A HSVPB failed to advance the subsequent atrial electrogram (*Figure 3B*). Considering the distance (55 mm) and acute angle between the medial aspect of the inferior caval vein (IVC) and the atrial septum created by right atrial anatomical

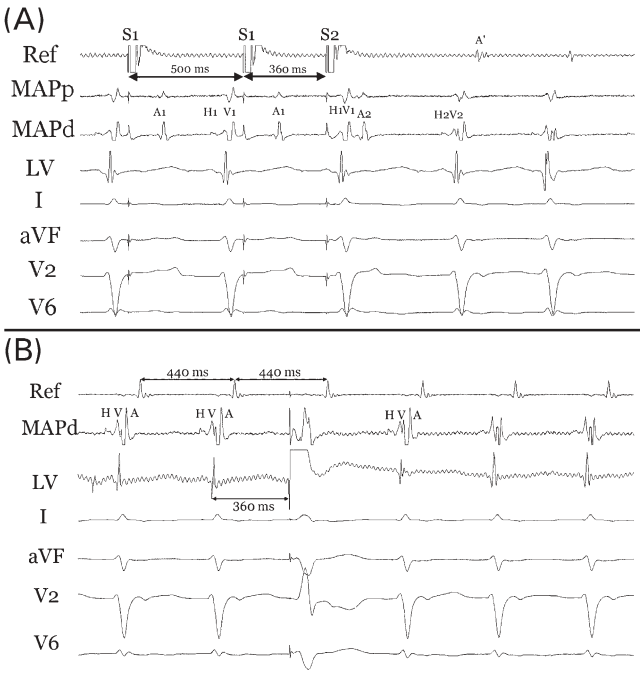

Figure 3 Surface electrograms from leads I, aVF, V2, and V6 with contact bipolar electrograms recorded from the right atrium (Ref), right atrial septum (map), and left ventricle (LV). (*A*) demonstrates programmed atrial stimulation (S1 500/S2 360) where the extra stimulus (S2) prolongs A2H2 leading to retrograde atrial activation (A') via the fast pathway (echo beat) and initiation of tachycardia. (*B*) During AVNRT (CL 440 ms) with the mapping catheter positioned approximating the His bundle a His (H) deflection can be seen leading ventricle (V) and atrium (A). Atrial activity was earliest in this area. A HSVPB at 360 ms captures the ventricle but fails to pre-excite the atrium supporting a diagnosis of AVNRT.

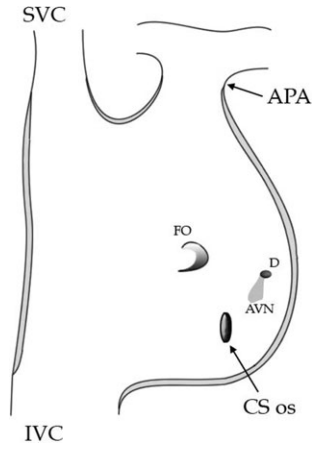

Figure 4 A drawing of the right atrium following the atriopulmonary Fontan procedure as performed in the patient described in this case viewed from the anterior approach. The position of the compact AVN is shown in the floor of the right atrium, anterior, and superior to the position of the CSos. The dimple (D) marks the position of the central fibrous body, the site of penetration of the His bundle. FO = fossa ovalis.

distortion, a RAMP sheath (St Jude Medical, St Paul, MN, USA) was used to improve stability of the right atrial ablation catheter and approach to the atrial septum. Radiofrequency energy limited to 50 W and 50°C was applied to the right side of the septum initially anterior to the ostium of the coronary sinus (CSos) and subsequently moving superiorly (*Figure 2B*) until an accelerated junctional rhythm was observed during ablation. Proximity of the ablation catheter to the His bundle was monitored on the electroanatomic map and from the left atrial catheter. Prolonged testing in the presence of atropine and isoprenaline failed to re-induce tachycardia, with no other arrhythmia inducible. There was no evidence of prolongation of the AH interval during sinus rhythm following ablation. The patient remains well on no anti-arrhythmic medication 4 months later with no symptomatic arrhythmia recurrence. Asymptomatic AVNRT recurrence has been excluded on 7-day Holter assessment.

Discussion

While different arrhythmia mechanisms have been documented following the atriopulmonary Fontan including both macro and micro-reentry,[1] we believe this to be the first description of AVNRT. Successful ablation of AVNRT has been described in a patient with tricuspid atresia palliated with a systemic to pulmonary shunt,[2] although in this setting the right atrium may not endure the same degree of dilation as following an atriopulmonary connection.

The diagnosis of AVNRT was based on the following findings: (i) atrial activation pattern during arrhythmia defined by electroanatomic mapping; (ii) reproducible initiation and termination of tachycardia with programmed stimulation; and (iii) the presence of atrial echo beats during programmed atrial stimulation indicative of dual AV nodal physiology. We could not demonstrate an AH jump according to standard criteria (AH prolongation of >50 ms with a 10 ms decrease in S1–S2), which may be due to the right atrial anatomical and physiological abnormalities, although this is frequently the case in young patients.[3] Atrial tachycardia was felt to be highly unlikely in the face of reproducible arrhythmia termination with antegrade AV block. Failure of an appropriately timed HSVPB to advance retrograde atrial activation excludes a para-Hisian accessory pathway.[4] Junctional tachycardia with retrograde conduction seems unlikely in the face of regular tachycardia cycle length inducible by programmed atrial stimulation, and the absence of capture beats or VA dissociation.[5]

In this patient, access to the right ventricle and constant and reliable recording of a His potential were prevented by the atretic tricuspid valve and the distance from the IVC (60 mm). We therefore elected to use a modified four-catheter technique to allow constant mapping of the right atrium, His bundle from the left atrial aspect, and left ventricle during ablation. In the event of atrioventricular block, temporary pacing could have been performed via the left ventricular catheter. The anatomical position of the AV conduction system has been well defined in tricuspid atresia. The compact AV node lies in the muscular floor of the right atrium adjacent to the atrial septum and inferior to the insertion of the tendon of Todaro and extends anteriorly to the central fibrous body, commonly marked by a dimple[6] (*Figure 4*). This is clearly different from the structurally normal heart, where the compact node lies in the apex of the triangle of Koch with clearly defined anatomical

boundaries, and the slow pathway is located within the septal isthmus between the tricuspid annulus and the CSos.[7] Application of radiofrequency energy adjacent to the CSos failed to elicit a junctional response and tachycardia remained inducible. Successful slow pathway ablation was finally achieved in a relatively superior position, close, but inferior to the location of the His bundle as defined by the left atrial catheter and tagged on the right atrial endocardium during sinus rhythm mapping (*Figure 2B*). Although cryoablation would eliminate the problems of catheter instability during ablation, it is not registered by the CARTO system and therefore was considered an inappropriate ablative modality considering the anatomical complexity and benefits of electroanatomic mapping.

Ablation of AVNRT after an intra-cardiac or extra-cardiac total cavopulmonary connection would be possible from the right side of the inter-atrial septum only in the presence of a fenestration between the 'systemic' and 'pulmonary' atria. In the absence of such a communication, access to the slow pathway would only be possible using a retrograde approach via the aorta and left ventricle, as demonstrated in this case, and ablation performed from the left side of the septum as has been successfully performed in a patient with tricuspid atresia and atrial septal defect.[2]

In summary, we describe successful ablation of symptomatic and recurrent AVNRT in a patient with an atriopulmonary Fontan, highlighting the importance of carefully evaluating arrhythmia mechanism in these patients. Potential difficulties of access and catheter stability were addressed using a combined antegrade and retrograde four-catheter approach with detailed bi-atrial mapping.

Acknowledgements

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