Clinical problem solving: Maneuvering around a narrow complex tachycardia in a patient with Mustard repair for transposition of the great arteries

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Introduction

A 43-year-old man with previous atrial switch repair for dextro-transposition of the great arteries (d-TGA) presented with recurrent palpitations associated with dyspnea and presyncope.

Case report

The patient was initially palliated with balloon atrial septostomy before undergoing Mustard repair at age 1. The subsequent course was complicated by both inferior vena cava baffle stenosis and obstruction to pulmonary venous flow requiring surgical correction. Following these procedures, a dual-chamber permanent pacemaker was implanted for bradycardia at age 22 owing to presumed sinus node dysfunction. Echocardiography confirmed satisfactory systemic (morphologic right) ventricular function with ventriculoarterial discordance noted. There was no evidence of recurrent baffle obstruction. Echocardiographic imaging was consistent with an excellent hemodynamic result from an atrial switch repair for d-TGA.

Owing to ongoing symptoms despite sotalol therapy, the patient was referred for an electrophysiology (EP) study. Catheter positions at EP study are shown in Figure 1A. The decapolar catheter was positioned in the systemic atrial appendage because the coronary sinus could not be identified and often empties into the pulmonary venous atrium (detailed surgical notes were not available to confirm this). Figure 1B shows a permanent pacemaker rhythm strip from a typical tachycardia episode (rate 180 beats/min). The electrocardiogram (ECG) during sinus rhythm is shown in Figure 1C and tachycardia induction with an atrial extrastimulus is shown in Figure 1D.

What is the most likely diagnosis for his tachycardia and how can this be proven with EP maneuvers?

Discussion

Inspection of the patient's tachycardia on the rhythm strip (Figure 1B) shows initiation by a premature ventricular complex and then 1:1 AV association follows with a different morphology on the ventricular electrogram compared with that seen in sinus rhythm. On further rhythm strips we saw this tachycardia terminate spontaneously with an atrial event as well. Initiation following a premature ventricular complex and termination with an atrial event makes atrial tachycardia unlikely.

Ventricular tachycardia (VT) may occur in patients following the Mustard procedure; however, it is much less common than atrial arrhythmias and sinus node dysfunction.[1,2] Figure 1D shows an example of the tachycardia with a relatively narrow morphology that is not dissimilar to the morphology in sinus rhythm (Figure 1C). Here it is also initiated with atrial pacing, which, combined with the relatively narrow morphology, makes VT unlikely, save for the possibility of fascicular VT. Pacing from the distal decapolar catheter (the systemic venous atrial appendage) at the start of Figure 1D demonstrates a broad QRS morphology with a slurred upstroke. With a left-sided accessory pathway, pacing here will demonstrate greater pre-excitation than during sinus rhythm (owing to proximity to the pathway). Following an atrial extrastimulus there is block in this pathway and delay in the AV node, which then allows orthodromic AV reentrant tachycardia (AVRT) to ensue.

How do we prove, however, that this is not atypical AV nodal reentrant tachycardia (AVNRT), atrial tachycardia, or fascicular VT where a left-sided bypass tract coincidentally blocks at initiation but does not take part in the tachycardia?

KEYWORDS Supraventricular tachycardia; d-TGA; Mustard repair; Electrophysiology ablation; Congenital heart disease; Wolff-Parkinson-White syndrome (Heart Rhythm Case Reports 2016;2:536–539)

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KEY TEACHING POINTS

- Owing to atrial dilatation and fibrosis following surgical incisions and baffle creation, intra-atrial reentrant tachycardia is by far the most common arrhythmia seen following Mustard repair for dextro-transposition of the great arteries (d-TGA).
- Typical AV nodal reentrant tachycardia has been reported following Mustard repair and, although accessory pathways are more common in patients with congenitally corrected transposition of the great arteries and Ebstein's anomaly, this case shows AV reentry tachycardia is possible in a patient with d-TGA.
- Advancing atrial activation during supraventricular tachycardia when the His bundle is refractory proves the presence of an accessory pathway. Other observations/maneuvers such as the change in VA interval with ipsilateral bundle branch block and the capacity to entrain the tachycardia from the site of the pathway with a short post-pacing interval help to prove accessory pathway participation and define the target for ablation.

In Figure 2A we see overdrive pacing from the left ventricular apex during tachycardia. Atrial activation is advanced with the first paced beat that captures the ventricle (the third QRS complex) and we can see from the surface ECG morphology that this is a fusion beat. Owing to proximity to the pathway, it is easier to advance atrial activation over a left-sided pathway when pacing from a left-sided subpulmonary ventricle than from the systemic ventricle. The capacity to reset the tachycardia with fusion proves the presence of an accessory pathway and tachycardia subsequently terminates.

We were unable to record a His bundle electrogram during tachycardia with a retrograde approach via the aorta and right ventricle to access the pulmonary venous atrium. While attempting to record a His signal, however, we noted the tachycardia spontaneously change from left bundle branch block to narrow complex and then to a right bundle branch block (RBBB) morphology (Figure 2B). The change in VA interval from 192 ms with left bundle branch block morphology to 122 ms and 102 ms with narrow complex and RBBB morphologies, respectively, demonstrates Coumell's sign and is consistent with orthodromic AVRT using a left-sided bypass tract. The fact that the VA interval is shorter with the RBBB morphology than with the narrow complex transition beat suggests that there is a degree of balanced delay in the left and right bundle branches, causing a narrow QRS complex as the left bundle branch recovers conduction and we revert back to the RBBB morphology seen at other times with tachycardia. This also demonstrates that the RBBB morphology seen on the sinus rhythm ECG relates to delay in the right bundle branch and hypertrophy of the right ventricle and not a complete, fixed conduction block in the right bundle branch.

Finally, in Figure 2C we see entrainment from the atrial aspect of the lateral mitral annulus during tachycardia. Tachycardia is accelerated to the paced rate and the post-pacing interval minus the tachycardia cycle length from the lateral mitral annulus is 20 ms. This further proves that the lateral mitral annulus is part of the circuit and confirms this tachyarrhythmia is not AVNRT with a bystander accessory pathway or fascicular tachycardia. We then went on to map the earliest atrial activation along the mitral annulus in tachycardia (Figure 2D) using the CARTO mapping system (Biosense Webster, Diamond Bar, CA). Earliest activation was identified at 2 o'clock on the mitral annulus and ablation here terminated tachycardia. A consolidation lesion was then performed just superior and inferior to this point (Figure 2D).

To our knowledge, this is the first reported case of successful ablation for AVRT in a patient with d-TGA and a Mustard repair. Greene et al[3] have previously reported a case series of AVNRT in this setting, and we carefully excluded this differential with the use of pacing maneuvers. Owing to atrial dilatation and fibrosis following

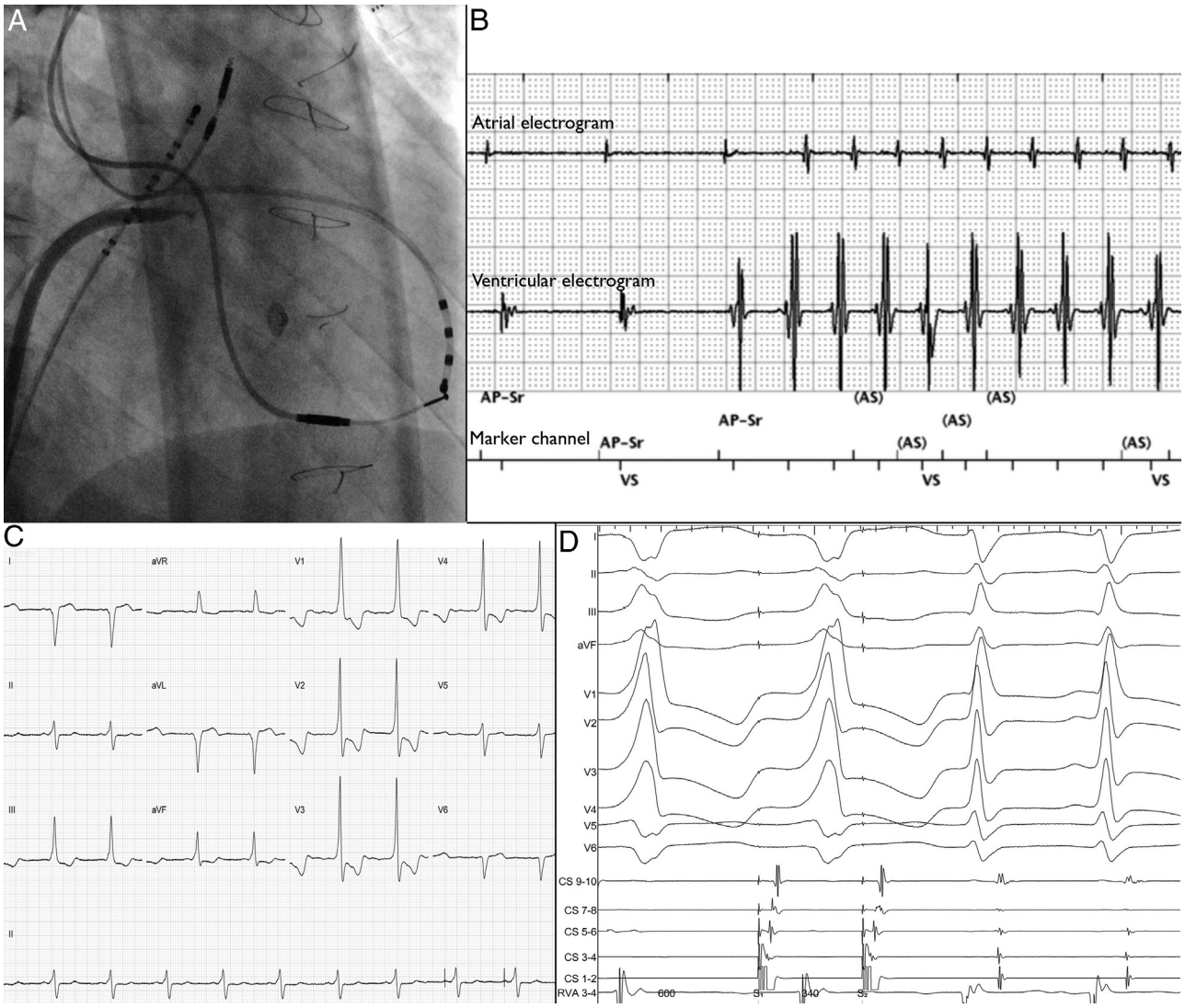

Figure 1 **A:** Fluoroscopic image in right anterior oblique that shows 2 pacing leads enter the heart through the superior vena cava baffle. The atrial lead sits in the systemic venous atrial appendage and the ventricular lead sits in the subpulmonic left ventricle. Decapolar and quadripolar electrophysiology catheters sit adjacent to these leads. **B:** Onset of a tachycardia episode from the patient's permanent pacemaker. The atrial and ventricular bipolar electrograms are shown along with the marker channel. **C:** Electrocardiogram (ECG) in sinus rhythm with right axis deviation and evidence of right ventricular hypertrophy (typical of a patient post Mustard repair). There is also a relatively short PR interval (130 ms) and a slurred onset to the QRS, which raises the suspicion of pre-excitation. **D:** Twelve surface ECG leads above atrial (CS 1-10) and ventricular (right ventricular apical) electrograms where an atrial extrastimulus initiates tachycardia.

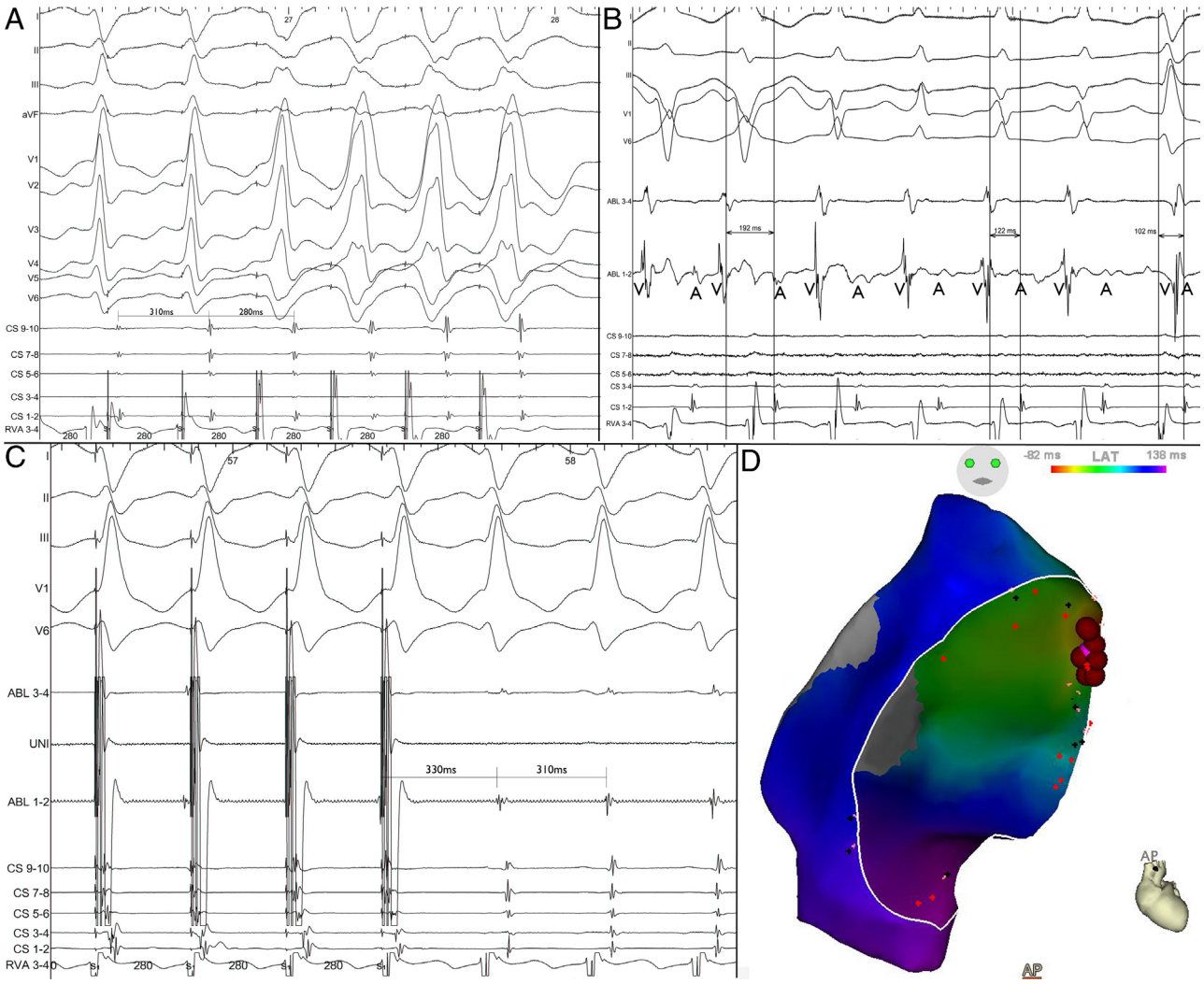

Figure 2 **A:** Overdrive pacing from the left ventricular apex (sub-pulmonic ventricle) during tachycardia. The decapolar catheter is positioned as shown in Figure 1A (as is the case in all panels). **B:** Variation in the VA timing during tachycardia when the QRS morphology changes from left bundle branch block to narrow complex to right bundle branch block morphologies (V denotes ventricular electrograms and A denotes atrial electrograms on the distal ablation catheter). **C:** Entrainment of tachycardia from the ablation catheter, which is sitting at 2 o'clock on the mitral annulus. The tachycardia cycle length and post-pacing intervals are shown. **D:** Electroanatomic map of the mitral annulus using the CARTO mapping system, where the timing of atrial activation was annotated during tachycardia. Earliest activation is shown in red and ablation lesions are shown as red dots over this point of earliest atrial activation (the accessory pathway location).

surgical incisions and baffle creation, intra-atrial reentrant tachycardia (IART) is by far the most common arrhythmia seen in patients following Mustard procedures.[2] Up to half of these IARTs are cavotricuspid isthmus dependent[4] and their presence confers an adverse outcome.[1] Accessory pathways are more frequently encountered in patients with congenitally corrected transposition of the great arteries (l-TGA) and Ebstein's anomaly,[5] with this association not typically described in d-TGA.

The patient has not had any arrhythmia recurrence in over 6 months of follow-up following sotalol withdrawal. Hence accessory pathway and AVRT recurrence are very unlikely at this stage. In contrast, even after successful ablation of IART, the long-term recurrence rates are very high owing to the underlying anatomic and electrophysiological substrate.[4] This case highlights the importance of considering different arrhythmia mechanisms in complex patient cohorts and the critical role of using EP maneuvers to confirm the correct diagnosis.

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