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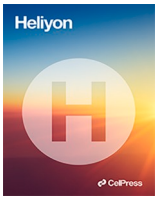

Case report

Case report: Antidromic atrioventricular reentrant tachycardia and underlying Ebstein anomaly

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A B S T R A C T

Multiple accessory pathways (APs) can develop in patients with Ebstein anomaly. Rarely, these APs can participate in antidromic atrioventricular reentrant tachycardia (AVRT) which can be life-threatening and requires unique considerations for acute management and ultimate ablation. These considerations are discussed herein.

1. Histroy of presentation

A 23-year-old woman without known medical history presented to the Emergency Department with palpitations, dyspnea, and intermittent chest tightness after consuming alcohol, multiple energy drinks, and cocaine. She was found to have a heart rate (HR) of 213 beats per minute, a blood pressure (BP) of 72/44 mmHg, and was awake and oriented but mildly lethargic. Electrocardiogram [EKG] (Fig. 1A) at presentation demonstrated a wide-complex tachycardia (WCT). She was given 1 mg of intravenous (IV) midazolam and cardioversion was performed with 100 J delivered via biphasic waveform. She was subsequently treated with a 150 mg IV bolus of amiodarone followed by a continuous infusion. Her HR improved to 82 beats per minute with a BP of 116/74 mmHg and repeat EKG is shown in Fig. 1B. Subsequent physical exam revealed a soft II/VI pansystolic murmur best appreciated over the right lower sternal border without radiation to the carotid arteries or back. Jugular venous pulsations corresponded to 7 cm of water and no pedal edema was appreciated.

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Abbreviations

1. HR heart rate
2. BP blood pressure
3. EKG electrocardiogram
4. WCT wide-complex tachycardia
5. IV intravenous
6. SVT supraventricular tachycardia
7. AVRT atrioventricular reentry tachycardia
8. AP accessory pathway
9. AV atrioventricular
10. AF atrial fibrillation
11. SPERRI shortest pre-excited R–R interval

2. Past medical history

The patient had no known prior medical history. She endorsed frequent lifelong palpitations occasionally accompanied by feelings of lightheadedness without prior syncopal episodes.

3. Differential diagnosis

The differential diagnosis for regular WCT includes ventricular tachycardia, supraventricular tachycardia (SVT) with aberrant intraventricular conduction, and pre-excited SVT including antidromic atrioventricular reentrant tachycardia (AVRT) via one or multiple accessory pathways (APs) [1]. Examination of the EKG in Fig. 1B revealed delta-waves and was thus suggestive of electrocardiographically manifest AP(s). In this instance, the AP(s) participated in antidromic AVRT mediated via one or multiple APs (such as in pathway-pathway dependent tachycardia). Antidromic AVRT occurs when anterograde conduction from atria to ventricles occurs via an AP with retrograde conduction occurring via the atrioventricular (AV) node (such as in Fig. 2) or another AP [1].

4. Investigations

The complete blood count and basic metabolic panel were normal with a positive urine toxicology screen for cocaine and marijuana. When AVRT is suspected evaluate for and address conditions that may precipitate/worsen reentrant rhythms (including ischemia, hyperthyroidism, thromboembolism, anemia, arrhythmogenic agents etc.) while also evaluating for structural cardiovascular disease on initial presentation. The patient had a negative high sensitivity troponin I level (<3 ng/L; normal: 0–34), a normal thyroid stimulating hormone level (0.756 uIU/mL; normal: 0.4–4.0), and an echocardiogram (Fig. 3) which was consistent with Ebstein anomaly or atrialization of a portion of her right ventricle with apical displacement of the tricuspid valve associated with severe tricuspid regurgitation and right atrial dilation.

5. Management

The patient underwent electrophysiologic mapping as shown in Fig. 4. Programmed electrical stimulation did not demonstrate evidence (eg echo beat) of dual AV node physiology. In the electrophysiology lab the patient presented in sinus rhythm with delta waves. A WCT was induced via ventricular extrastimuli (tachycardia cycle length 320 ms) and with ventricular overdrive pacing (at 290 ms) the atrial signal advanced within 2 beats. Once the tachycardia was entrained with a ventricular-atrial-ventricular response the post-pacing interval tachycardia cycle length was 10 ms and the difference between the stimulus-atrial interval and the ventriculoatrial interval was 17 ms. These maneuvers excluded atrial tachycardia and AV nodal reentrant tachycardia.

The arrhythmia deteriorated into atrial fibrillation (AF) and required multiple cardioversions and IV ibutilide (to continue mapping and ablation), thus demonstrating high-risk properties. In sinus rhythm the manifest AP localized to the anterolateral tricuspid annulus (at 10 o'clock) with similar localization with retrograde ventriculoatrial mapping (Fig. 4). Ventriculoatrial conduction occurred mainly via the anterolateral AP with ventricular pacing (Fig. 5A) as did atrioventricular conduction in normal sinus rhythm (Fig. 5B). During AVRT ventriculoatrial activation was also observed via the mid-septal AP (Fig. 5C). Unfortunately, ablation was complicated by severe right atrial enlargement and ablation of the right anterolateral AP was unsuccessful.

Subsequent attempts to induce tachycardia led to typical atrial flutter and thus the cavotriscupid isthmus was successfully targeted for ablation. This then allowed for AVRT to be reinduced with isoproterenol, long enough to map the retrograde limb to the mid-septal area, 8 mm from the bundle of His (Fig. 4). Radiofrequency ablation of the mid-septal AP was performed successfully. Consequently, AVRT could no longer be induced with aggressive atrial and ventricular stimulation (including with the provision of isoproterenol). Notably, the shortest pre-excited R–R interval (SPERRI) in atrial flutter and atrial fibrillation during the study was 307 ms.

Ultimately, this suggested that the AVRT was pathway-pathway dependent with anterograde conduction via the anterolateral AP and retrograde conduction via the mid-septal AP.

6. Discussion

When present, APs manifest pre-excitation on EKGs (e.g., a delta wave) in 70% of children, while the rest are concealed [1]. The prevalence of manifest APs in the general population is 0.1–0.3% [2]. Wolff Parkinson White Syndrome describes manifest ventricular pre-excitation in combination with SVT and can be associated with syncope or sudden death. In 90–95% of patients, these SVTs are orthodromic AVRT while approximately 5% are antidromic AVRT (Fig. 6) [3]. About 1.6% of patients with AVRT have structural cardiovascular disease [4]. Ebstein anomaly often leads to the development of multiple APs and requires special considerations in management [5]. In addition, the EKG aids in localizing APs (eg Arruda criteria, sensitivity 90%, specificity 99%) however, it should be

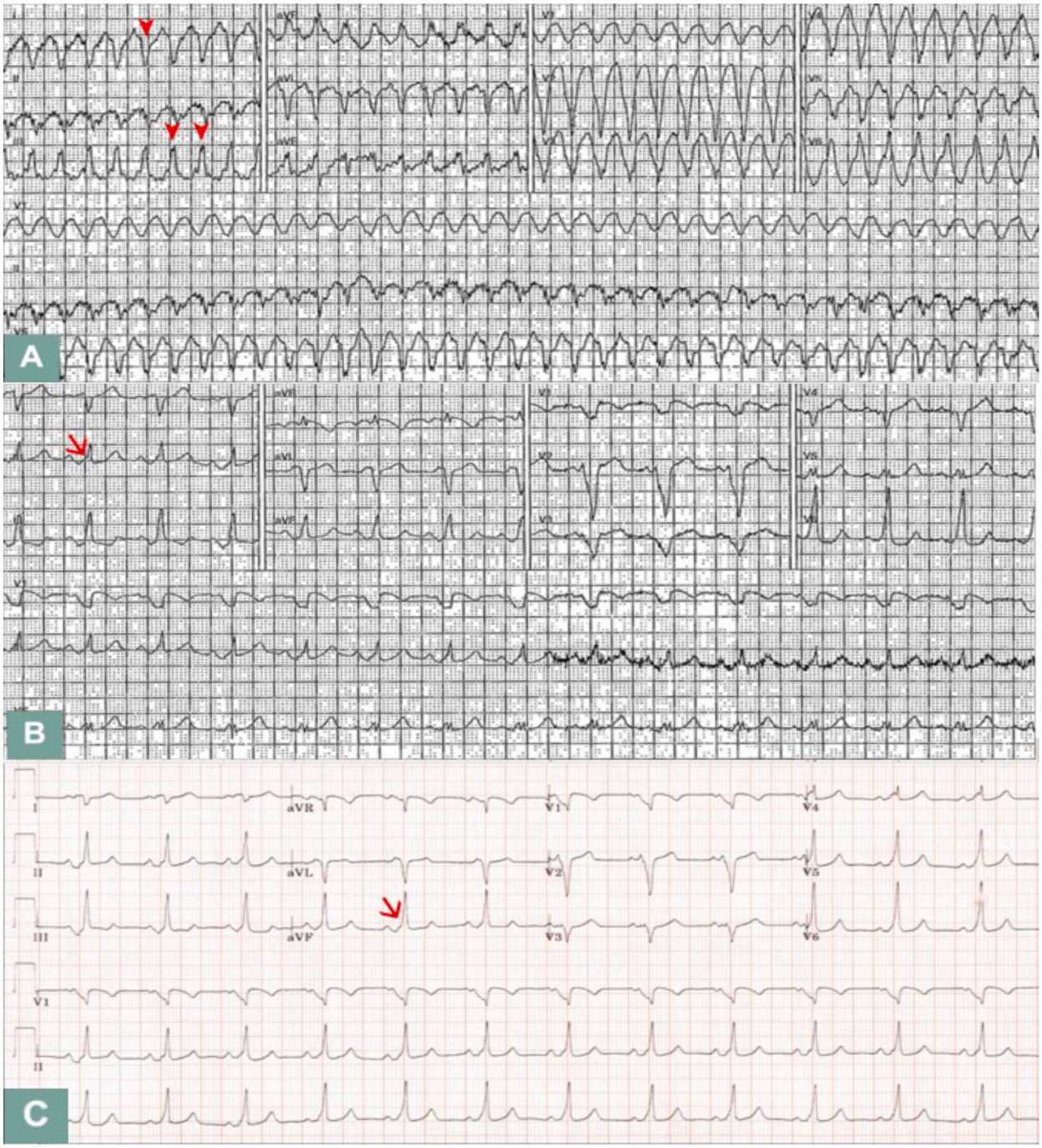

Fig. 1. Patient's EKGs. *Panel A*: EKG upon arrival with wide-complex tachycardia (arrowheads). *Panels B–C*: EKG after cardioversion (B), after ablation, and at hospital discharge (C) note shortened PR intervals and delta waves (arrows).

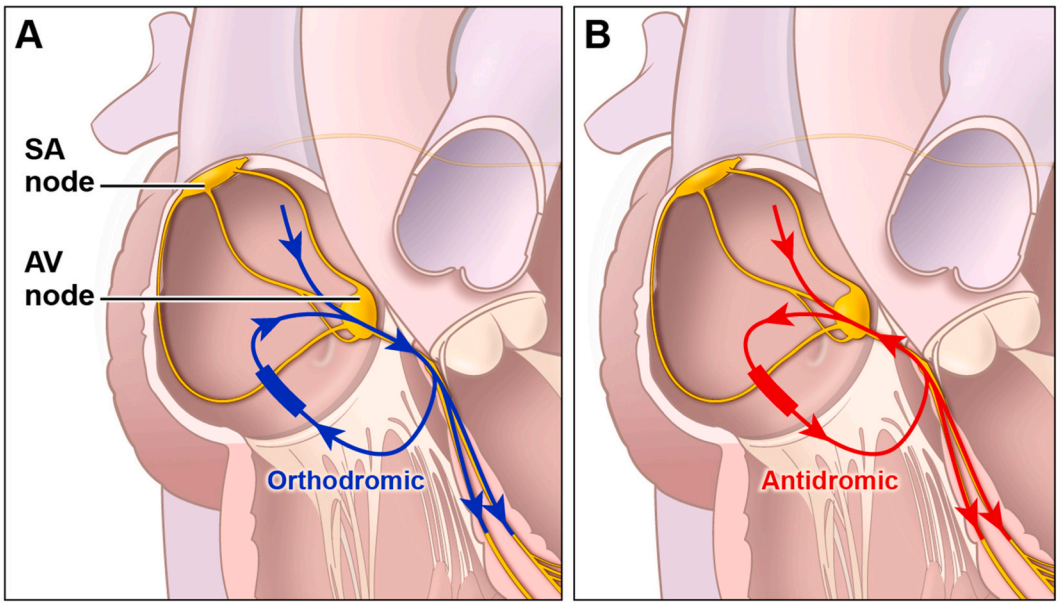

Fig. 2. AVRT Conduction Pathways. *Panel A*: orthodromic AVRT (QRS <120) arises from anterograde conduction via the atrioventricular (AV) node and retrograde via an accessory pathway (AP). *Panel B*: antidromic AVRT (QRS >120 ms) arises from anterograde conduction via the AP and retrograde via the AV node. Abbreviations: SA: sinoatrial.

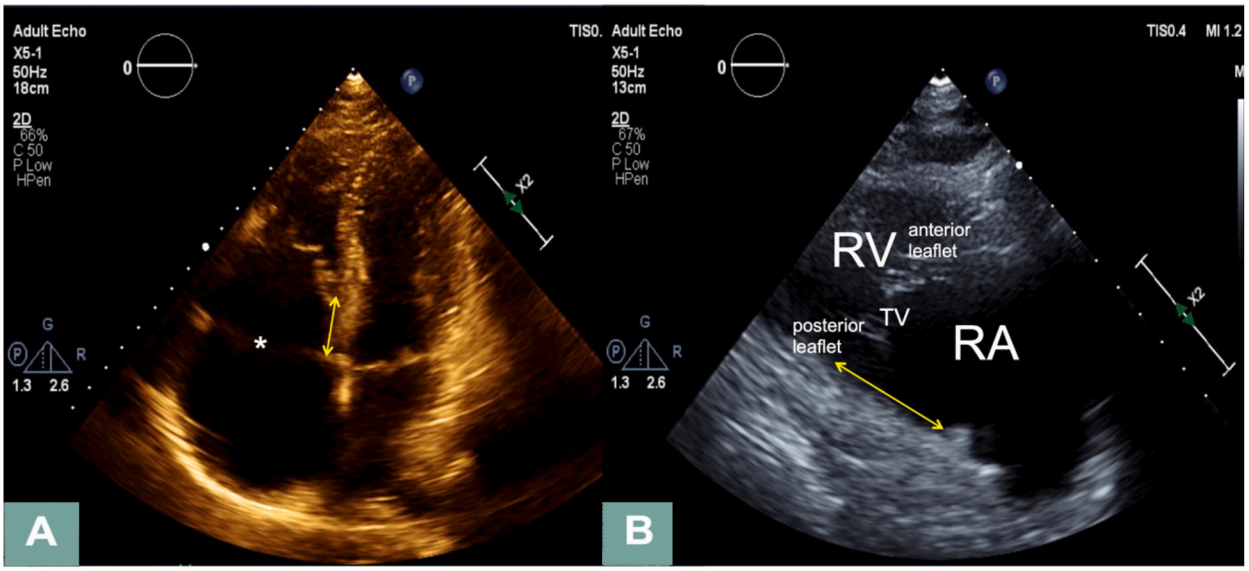

Fig. 3. Patient's Echocardiogram. *Panel A*: four-chamber view with typical findings of Ebstein anomaly with right atrial (RA) dilation and right ventricular (RV) atrialization (asterisk) of the tricuspid valve (TV) relative to the mitral annulus (double arrows). *Panel B*: RV inflow view demonstrating apical TV displacement (double arrows).

noted that patients with Ebstein anomaly may have distorted anatomy (such as in our patient, see Fig. 7) [6]. For example, in the setting of right ventricular strain and atrialization of the tricuspid valve (Fig. 7) the normal R-wave morphology appreciated in lead I may be opposite (as in Fig. 1) with a [less dominant] r-wave (corresponding to depolarization of a small right ventricle) and a [dominant] S-wave (corresponding to a more laterally displaced left ventricle). Thus, in light of potentially distorted anatomy, caution may be warranted when interpretting EKG patterns in patients with Ebstein anomaly. The malformed tricuspid valve and consequent tricuspid regurgitation in Ebstein anomaly also predisposes these patients to AF. This is a critical consideration as rapid conduction of AF to the ventricles, via an AP, can lead to sudden cardiac death (10-year risk of 0.15–0.24%) in patients with manifest APs [3]. Upon invasive electrophysiologic studies, the shortest pre-excited R–R interval, or SPERRI, less than 250 ms (in our patient 307 ms),

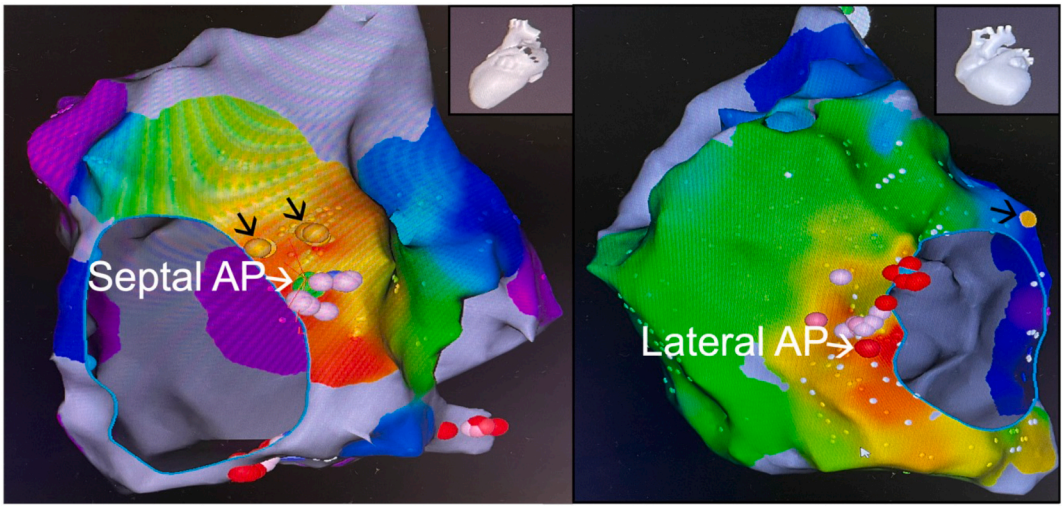

Fig. 4. Patient Three-Dimensional Electroanatomic Map Images. Three-dimensional electroanatomic mapping showing presence of dual right anterolateral and mid-septal APs (white arrows). The tachycardia terminated with ablation of the septal AP and was not subsequently re-inducible. The yellow dots mark the bundle of His (black arrows). (For interpretation of the references to colour in this figure legend, the reader is referred to the Web version of this article.)

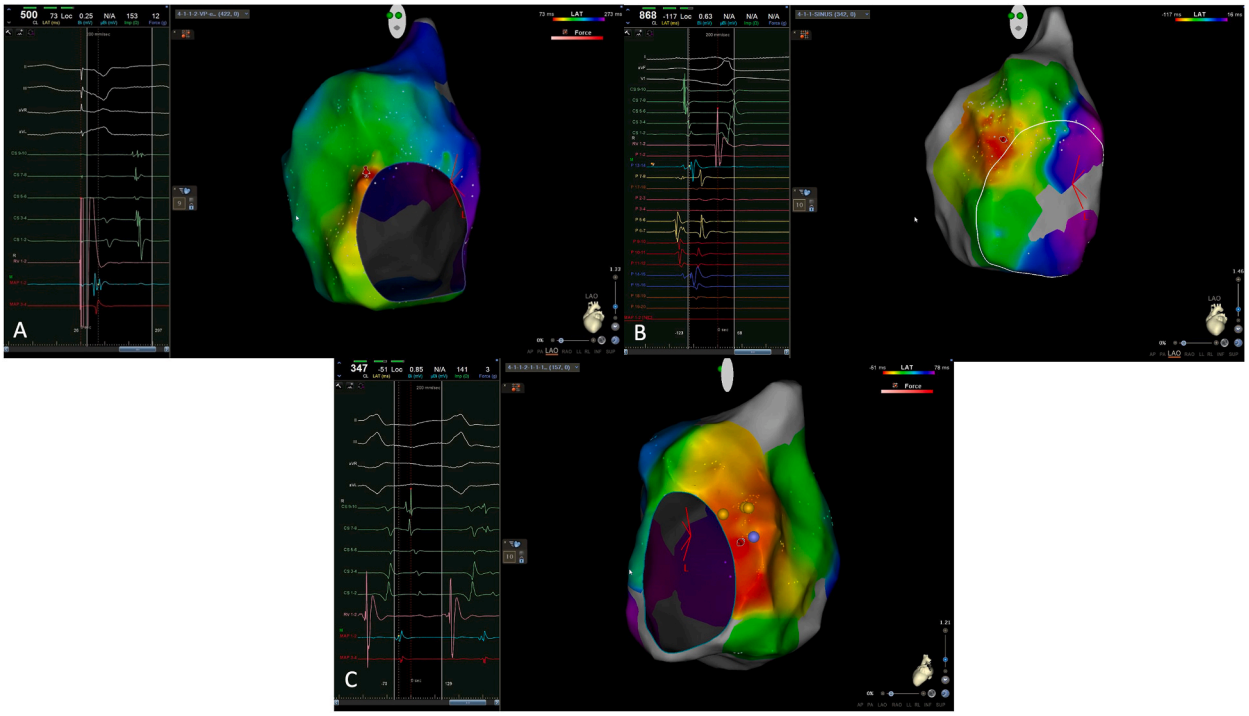

Fig. 5. Intracardiac Electrocardiograms. Note ventriculoatrial activation with ventricular pacing (*Panel A*) and atrioventricular activation* during sinus rhythm (*Panel B*), both via the (more dominant) anterolateral accessory pathway (AP) [as more readily appreciated on the corresponding intracardiac electrocardiograms]. *Panel C*: demonstrating ventriculoatrial activation via the (less dominant) mid-septal pathway during AVRT. *Note the activation map (Fig. 5B) demonstrating the earliest ventricular activation at the right anterolateral AP (red pin; also see Fig. 4). This site is located along the tricuspid annulus (white circle) which is also in the atrialized portion of the right ventricle. (For interpretation of the references to colour in this figure legend, the reader is referred to the Web version of this article.)

indicates that an AP has high-risk properties and may permit rapid AV conduction subsequently leading to cardiac arrest [7]. Given the presence of often multiple APs and the factors contributing to AF, patients with Ebstein anomaly may be at higher risk for sudden cardiac death.

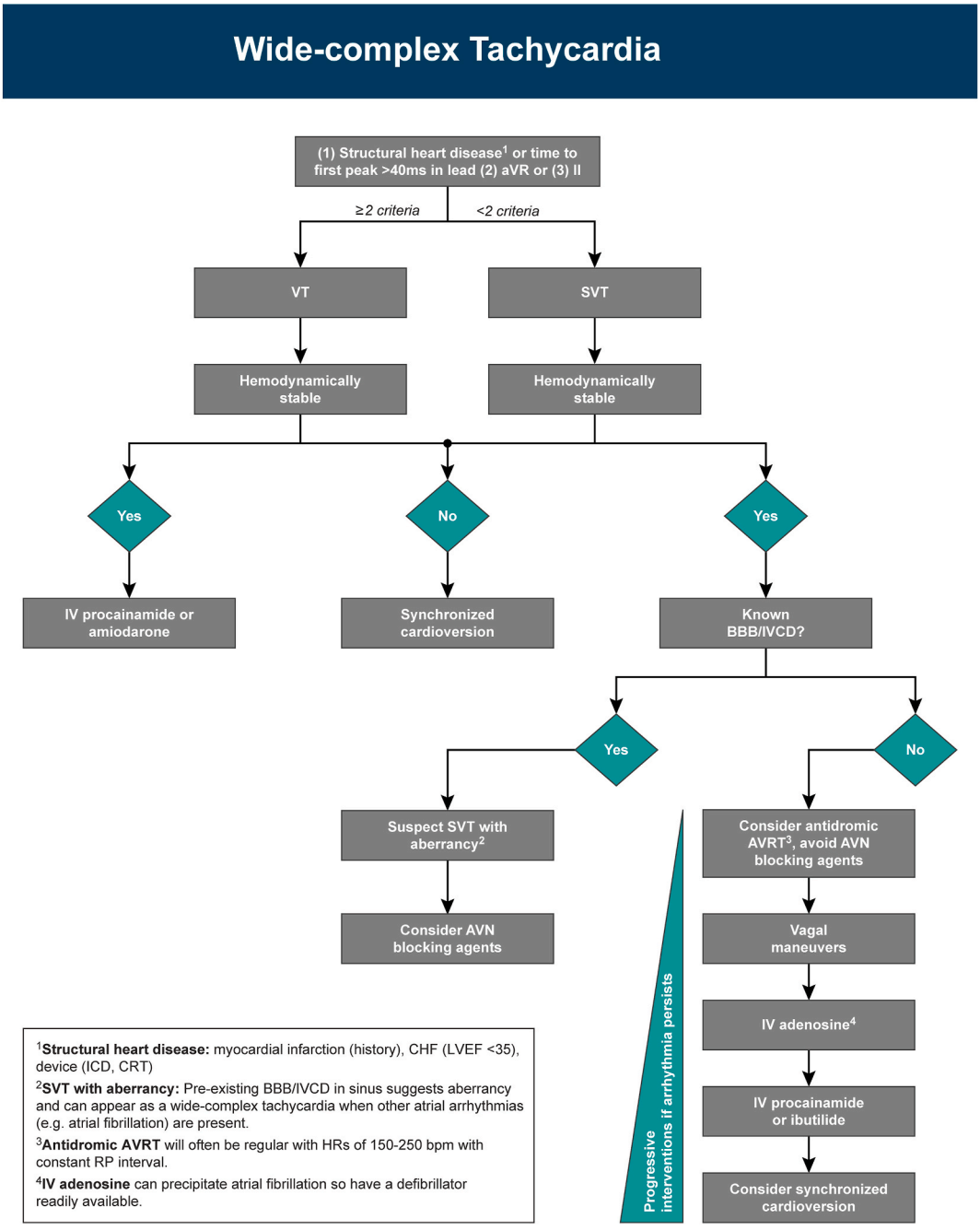

Fig. 6. Identification and Management of Wide-Complex Tachycardias and Antidromic AVRT. Note medications and dosing in Table 1. Abbreviations: AVN: atrioventricular nodal; AVRT: atrioventricular reentrant tachycardia; BBB: bundle branch block; IVCD: intraventricular conduction disturbance; SVT: supraventricular tachycardia; VT: ventricular tachycardia.

In hemodynamically unstable patients with WCT, cardioversion should be performed and advanced cardiac life support and pediatric advanced life support algorithms should be followed [8,9]. In hemodynamically stable patients that do not meet criteria for ventricular tachycardia, suspect SVT with aberrancy (Fig. 6). Management strategies for WCT may vary widely amongst institutions/providers, a suggested approach incorporating American Heart Association/American College of Cardiology guidelines are outlined in Fig. 6 with medication dosing listed in Table 1—these decisions should be made alongside a cardiologist/electrophysiologist [3]. Otherwise in patients with antidromic AVRT, vagal maneuvers may be attempted (27.7% effective) followed by IV adenosine (Class Ib, 90–95% effective) then IV procainamide or IV ibutilide (Class Ic) and synchronized cardioversion (Class Ib) if pharmacotherapy fails (Fig. 6) [3].

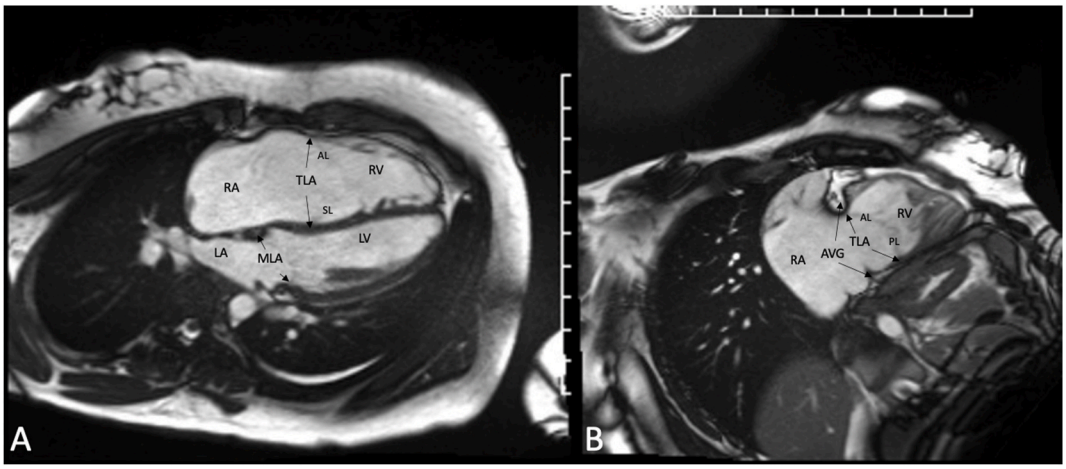

Fig. 7. Cardiac Magnetic Resonance Imaging: Distorted Anatomy. See bright blood cine images in four chamber (*Panel A*) and RV long axis (*Panel B*) views demonstrating distorted intracardiac anatomy with apical displacement of the tricuspid septal and posterior leaflet attachments leading to a severely enlarged right atrium and a small right ventricle. Abbreviations: AL: anterior leaflet; AVG: atrioventricular groove; LA: left atrium; LV: left ventricle; MLA: mitral leaflet attachments; PL: posterior leaflet; RA: right atrium; RV: right ventricle; SL: septal leaflet; TLA: tricuspid leaflet attachments.

Table 1
Medications and dosing.

Agent/Intervention	Dosing	Considerations
IV Adenosine	6 mg repeat with 12 mg if unsuccessful	•Administer as a rapid push medication •Avoid in patients with decompensated obstructive lung disease and heart failure
IV Diltiazem	15–20 mg bolus over 2 min every 15 min ×3 doses	•Beware of hypotension
IV Verapamil	5–10 mg over 3 min every 15 min ×3 doses	•Avoid in patients with heart failure
IV Procainamide	100 mg every 5 min (max 17 mg/kg) Followed by continuous infusion of 20–50 mg/min	•Beware of hypotension •Avoid in patients with heart failure •Monitor for QT prolongation
IV Ibutilide	<60 kg: 0.01 mg/kg (0.1 mL/kg) may repeat 1 dose in 10 min >60 kg: 1 mg, may repeat 1 dose in 10 min	•Monitor for QT prolongation when initiating treatment
IV Esmolol	500-1000 mcg/kg over 30 sec Followed by continuous infusion of 150–300 mcg/kg/min	•Avoid in patients with decompensated obstructive lung disease and heart failure
Synchronized Cardioversion	100 J then 200 J (mono or biphasic)	

Unfortunately, as previously mentioned, orthodromic or antidromic AVRT can degenerate into AF. This is because the AP may not possess the same decremental conduction properties as the AV node, potentially leading to 1:1 conduction of atrial rates to the ventricles leading to ventricular fibrillation and cardiac arrest [2,3]. IV verapamil/diltiazem and beta blockers, which can slow AV nodal conduction, can favor AV conduction via the AP [1,3]. Therefore, these agents should only be considered as first-line agents in patients without pre-excitation on resting EKGs (Class IIa), otherwise they should only be considered when patients have not responded to other therapies (Class IIb) and avoided in patients with AF [3]. Thus, until wide-complex SVTs can be further classified assume antidromic AVRT is present and avoid AV nodal blocking agents.

Definitive diagnosis of AVRT and management of symptomatic AVRT occurs in the electrophysiology laboratory with ablation. Antiarrhythmic agents can be considered in patients with infrequent/well-tolerated AVRT [2]. Asymptomatic patients have an approximate 0.3% risk over 5–20 years of ventricular fibrillation, therefore these patients should undergo electrophysiologic study with ablation if SVT is easily induced, or if the patient has a high-risk occupation (i.e. airline pilot) [2,3].

6.1. Back to the patient

In this patient, Ebstein anomaly (abnormal tricuspid valve development giving rise to multiple APs) with severe tricuspid regurgitation (placing a wall stress on the right atrium) and right atrial enlargement (chamber dilation) in the setting of sympathomimetic agents (namely cocaine shortening the refractory period of the AV node) provided the ideal arrhythmogenic environment to precipitate hemodynamically unstable AVRT. Without early intervention, these same conditions may have led to AF placing the patient at

risk of life-threatening ventricular fibrillation.

After cardioversion, IV amiodarone, and ultimate ablation of the mid-septal AP, the right anterolateral AP remained and manifest pre-excitation was noted on her EKG at discharge (Fig. 1C). Ablation of APs is often quite successful (92–98%), however, multiple APs are often present in patients with Ebstein anomaly requiring repeat ablations (odds ratio 2.5, 95% confidence interval 1.2–5.4) [4,10]. Ultimately, septal APs, given their proximity to the AV node can be challenging to successfully ablate and lead to recurrence of AVRT in about 10–15% of patients [10]. In this patient the anterolateral AP could not successfully be ablated due to atrial dilation—a consequence of longstanding tricuspid regurgitation and another consideration in patients with Ebstein anomaly. She continued to have pre-excitation on her EKG (a limitation of this case) and was offered the option for repeat mapping and ablation of the anterolateral AP, but due to insurance issues she had to seek care in another state.

6.2. Follow-up

After ablation a continuous 14 day extended Holter monitor was placed and demonstrated manifest pre-excitation without evidence of SVT.

7. Conclusions

Multiple APs can form in patients with Ebstein anomaly and rarely present with antidromic AVRT. There is a higher likelihood of persistent APs with need for re-ablation in patients with Ebstein anomaly given multiple APs. Though rare, sudden cardiac death may occur if rapid atrial rates (such as in the setting of AF) are conducted via APs to the ventricles as this leads to ventricular fibrillation.

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Declarations

This work has not been published previously and is not under consideration for publication elsewhere. The patient consented to this publication which is approved by all authors and responsible parties.

CRediT authorship contribution statement

Matthew A. Nazari: Writing – review & editing, Writing – original draft, Supervision, Project administration, Methodology, Investigation, Funding acquisition, Conceptualization. **Natalie Dapas:** Writing – review & editing, Writing – original draft, Methodology, Investigation, Conceptualization. **Karel Pacak:** Writing – review & editing, Writing – original draft, Investigation, Funding acquisition, Conceptualization. **Zhengping Zhuang:** Writing – review & editing, Writing – original draft, Funding acquisition, Conceptualization. **Jared S. Rosenblum:** Writing – review & editing, Writing – original draft. **Abhishek Jha:** Writing – review & editing, Writing – original draft. **Arooge Towheed:** Writing – review & editing, Writing – original draft, Conceptualization. **Mark C. Haigney:** Writing – review & editing, Writing – original draft, Investigation, Conceptualization. **Athanasios Thomaides:** Writing – review & editing, Writing – original draft, Methodology, Investigation, Conceptualization, Supervision. **Monvadi B. Srichai:** Writing – review & editing, Writing – original draft, Supervision, Methodology, Investigation, Conceptualization.

Declaration of competing interest

The authors declare the following financial interests/personal relationships which may be considered as potential competing interests: Matthew A. Nazari reports financial support was provided by the National Institutes of Health.

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