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# Indian Pacing and Electrophysiology Journal

journal homepage: [www.elsevier.com/locate/IPEJ](http://www.elsevier.com/locate/IPEJ)

## Right atrial appendage tachycardia: A rare cause of tachycardia induced cardiomyopathy in a 4-year-old child



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### ARTICLE INFO

#### Article history:

Received 16 May 2018

Received in revised form

4 June 2018

Accepted 3 July 2018

Available online 4 July 2018

#### Keywords:

Tachycardiomyopathy

Right atrial appendage

Atrial tachycardia

Pediatric arrhythmia

Radiofrequency ablation

### ABSTRACT

We present a rare case of tachycardiomyopathy in a 4-year-old girl. The child had incessant atrial tachycardia (AT) and refractory heart failure. Right atrial appendage (RAA) was localised as the source of the ectopic tachycardia. The child underwent successful radiofrequency ablation (RFA) using 3-D electroanatomical mapping. Fluoroscopy was used sparingly only to rule out underlying anomalies. The left ventricular functions returned to normal by one month after the procedure. RAA AT is rare in very young children and usually necessitates surgical appendectomies. RFA is a challenge in such age groups and there are very few published literature on RAA AT in very young children.

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### 1. Introduction

Pediatric arrhythmias leading to tachycardiomyopathy (TCMP) are usually incessant ectopic atrial tachycardias (ATs), persistent junctional reentrant tachycardias (PJRT) and atrial flutters [1]. Right atrial appendage (RAA) ATs constitute 0.6–8% of all ectopic atrial tachycardias [2]. These forms of ATs are usually incessant and are known to cause TCMP in older children and in adults [3]. However, it is unusual that ectopic ATs originate from the RAA in very young children and there are only anecdotal reports of the same. We describe a rare case of right atrial appendage tachycardia leading to TCMP in a 4-year-old child.

### 2. Case report

A 4-year-old girl from Kenya, weighing 18 kg and with a height of 104 cm, presented to us with clinical features of heart failure. The mother of the baby also complained of persistent pounding of the

precordium in the baby since the last 6 months. The baby was already being treated for heart failure with diuretics, digoxin and beta blockers. Transthoracic echocardiography (Supplementary video 1) showed a dilated heart and severe left ventricular dysfunction. Obvious anatomical causes of left ventricular dysfunction were ruled out by trans-thoracic echocardiography. The 12-lead ECG showed a regular narrow QRS tachycardia with a very rapid ventricular rate of 280 beats per minute with a RP interval of 90 ms and upright P waves in the inferior leads (Fig. 1A). Adenosine, given during the tachycardia, resulted in continuation of the tachycardia with transient atrio-ventricular block (AV) block seen as rapid P waves without QRS complexes. The differential diagnoses were atrial tachycardia with 1:1 atrio-ventricular conduction and a clockwise atrial flutter. As the drugs were ineffective in controlling the arrhythmia and the heart failure, radiofrequency ablation was considered.

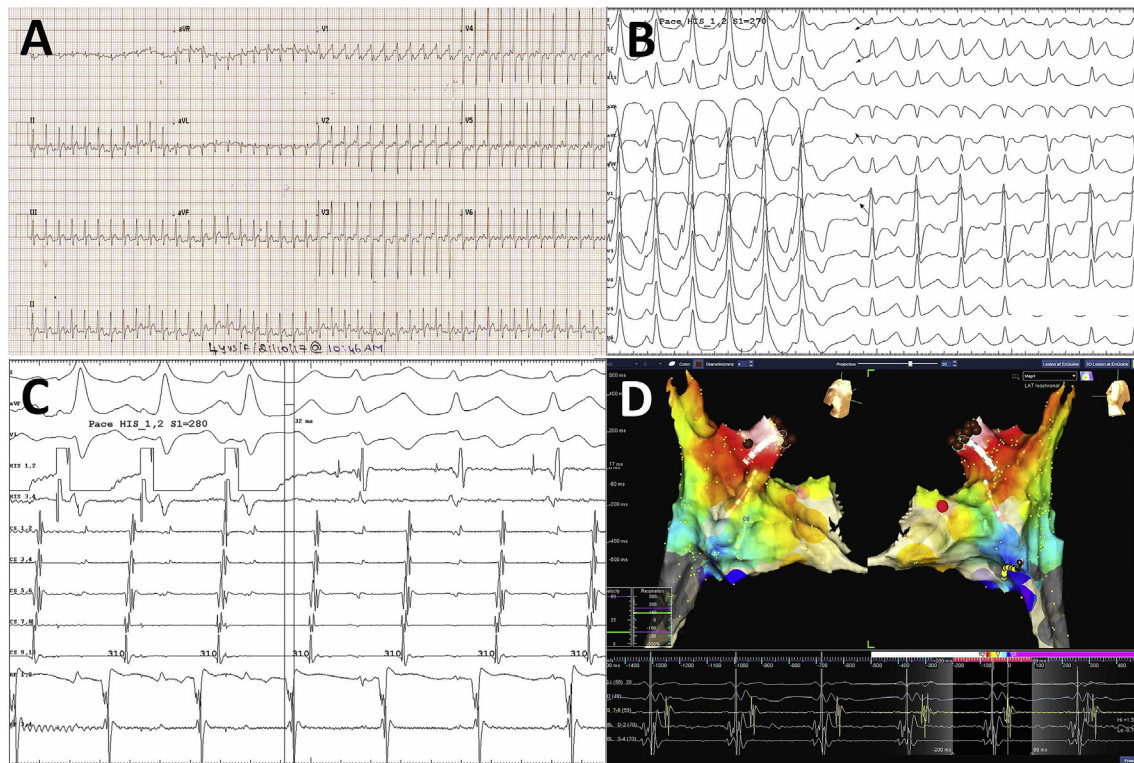
Supplementary video related to this article can be found at <https://doi.org/10.1016/j.ipej.2018.07.001>.

The baby was electively intubated for the procedure. She was in persistent tachycardia since the start of the procedure. Three venous accesses were obtained. A quadri-polar catheter (2 mm inter-electrode spacing) was used to map the His-bundle. A deca-polar catheter (2-5-2-mm inter-electrode spacing) was placed in the coronary sinus (CS). Another deca-polar catheter (2-5-2 mm inter-

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Peer review under responsibility of Indian Heart Rhythm Society.



**Fig. 1.** (A) Shows a regular narrow QRS tachycardia noted in the 4-year-old child with features of heart failure; (B) shows the positive P waves in leads II, III (downward arrows) and negative P waves in leads aVR and V1 (upward arrow), also seen is the slow P wave progression across the chest leads. The ventricle was overdrive paced as seen in the first half of the ECG; (C) shows the intra-cardiac signals. The His catheter is being utilized for ventricular overdrive pacing. Atrial signals in the CS catheter show an atrial tachycardia cycle length of 310 m s. The atrial bipolar signals at the RF catheter, which is placed near the right atrial appendage (RAA), reveals early signals with respect to the onset of P wave on the surface ECG; (D) shows 3D signals and ablation of RA appendage tachycardia using NavX Precision system (St. Jude Medical, St. Paul, MN, USA). Isochronal activation map has been constructed during tachycardia using proximal CS atrial signal as a reference. The RAA and the right atrium is seen in the RAO view (first half) and the left lateral view (second half). The color-coded display of the activation time in the right atrium measured relative to the reference point with red being early and blue being late. Activation map shows a very early target (white) at the base of the right atrial appendage. Ablation sites are colored brown.

electrode spacing) was used to map the right atrium (RA). Three dimensional (3-D) electro-anatomical mapping using NavX Precision system (St. Jude Medical, St. Paul, MN, USA) was utilized for mapping of the tachycardia. Radiofrequency ablation (RFA) was performed using an open-irrigated, 7F, 4 mm tip, quadri-polar catheter (Coolflex, St. Jude). No fluoroscopy was used for placement of catheters or for mapping of the atrium or even for the ablation.

Intra-cardiac signals revealed an atrial cycle length (CL) of 310 m s (Fig. 1C). A 1:1 atrio-ventricular conduction resulted in ventricular signals of the same CL. The V-V intervals (Fig. 1C) showed subtle variations due to changing A-H intervals. The activation time of bilateral atria from the RA free wall to CS- distal was only 40 m s. Entrainment from the different sites of the tricuspid annulus, the proximal and distal CS electrodes resulted in overdrive suppression and reinitiation of the tachycardia at long post-pacing intervals (390–480 m s) with variable V-A linking. All these evidences had effectively ruled out a reentrant mechanism of the tachyarrhythmia.

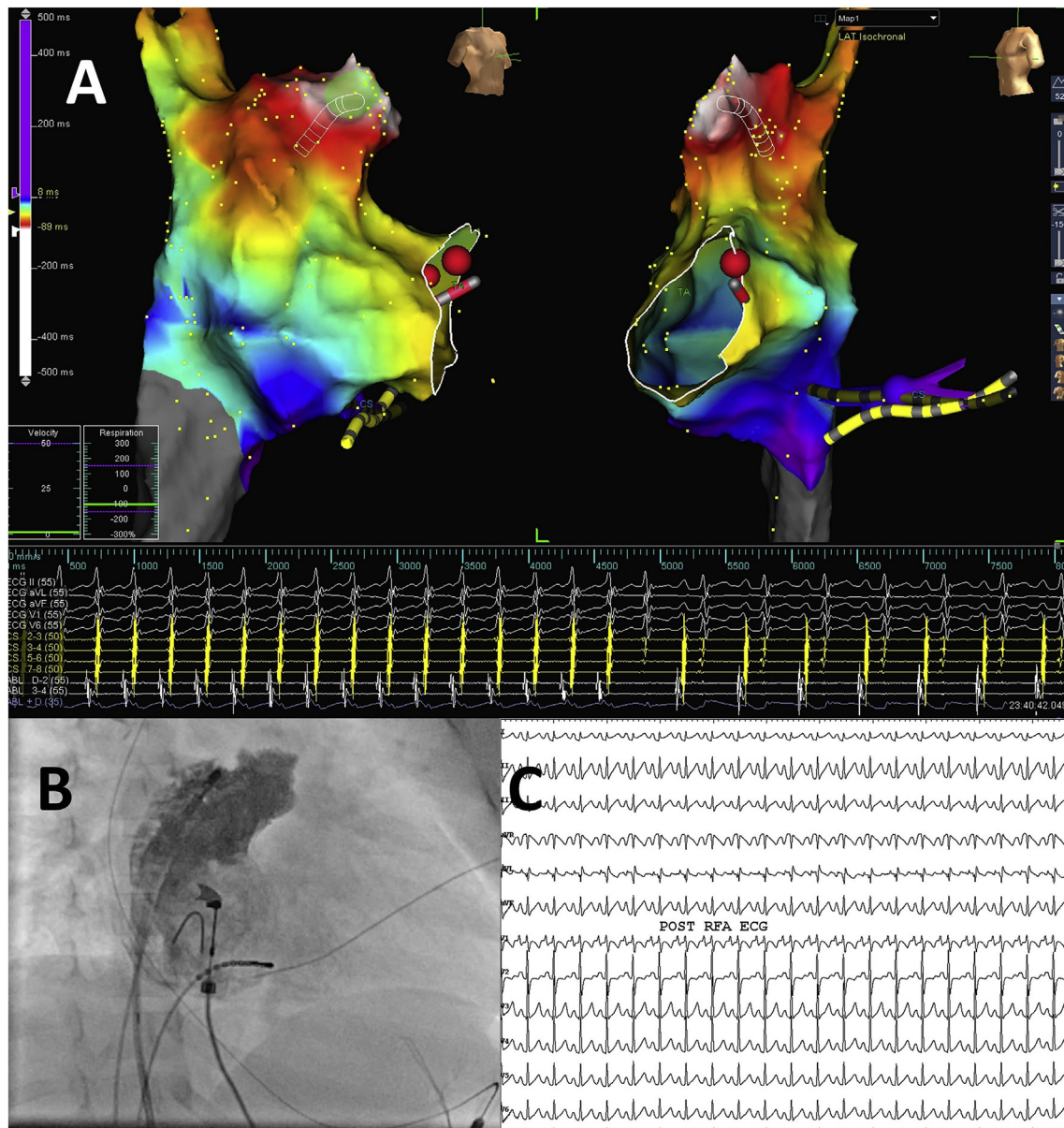
Ventricular overdrive pacing resulted in suppression and reinitiation of the tachycardia with an ectopic P wave (Fig. 1B). The P wave morphology was studied to help in locating the ectopic origin of the atrial tachycardia. Positive deflections in the inferior leads, negative deflection in V1 and progressive increase in p-wave amplitude across the chest leads suggested a right atrial appendage or crista terminalis (CT) or a superior tricuspid annulus as the probable origin of the tachycardia. Mapping near the RA appendage (RAA) showed early signals in the region of RAA. The 3-D electro-anatomical map suggested a centrifugal pattern of right atrial

activation spreading from the base of the RAA (Supplementary video 3). The His-signals were tagged (Fig. 1D).

Supplementary video related to this article can be found at <https://doi.org/10.1016/j.ipej.2018.07.001>.

Further mapping was carried out using the ablation catheter. The base of the RAA showed the earliest atrial activation signals (−32 m s) with respect to the onset of P wave on the surface ECG (Fig. 1C). Pacing at this site also resulted in short post-pacing intervals (330 m s). RFA was performed targeting a power of 25 watts (W), temperature of 40° C, lasting for 10–20 seconds using an open-irrigation catheter with a flow rate of 17ml/minute. After 2–3 lesions, the power was uptitrated to 30 W. Within 10 seconds of the RF burn the tachycardia terminated after initial prolongation of cycle length (Fig. 2A), after which the burn was continued for 60 seconds. At the end of the procedure, an angiogram of the RAA had ruled out anomalies of the appendage (Fig. 2B). The 12-lead ECG, after ablation, revealed a sinus origin of the rhythm (Fig. 2C) which was eventually confirmed by recording the atrial activation pattern at the high RA. Acute procedural success was achieved, as defined by the inability to induce tachycardia 30 min after ablation despite aggressive burst atrial pacing and the use of isoproterenol. The baby was extubated on the same day. The left ventricular functions had returned to normal by the fourth week of the procedure (Supplementary video 2). The baby was discharged without any cardiac medications. The baby was followed-up at 9 months after the discharge and there was no recurrence.

Supplementary video related to this article can be found at <https://doi.org/10.1016/j.ipej.2018.07.001>.



**Fig. 2.** (A) Shows termination 3D mapping and ablation of RA appendage tachycardia using NavX Precision system (St. Jude Medical, St. Paul, MN, USA). The termination of the atrial tachycardia was at the base of the right atrial appendage; (B) shows the angiogram of right atrial appendage using a pig-tail catheter in the RAO projection ruling out aneurysms and the position of the ablation catheter denotes the site of successful ablation at the superior base of the RAA; (C) 12-lead ECG shows the return of sinus rhythm after the ablation.

### 3. Discussion

We have described a rare case of incessant right atrial appendage tachycardia leading to TCMP in a 4-year-old child. ATs resulting in TCMP, though not uncommon in children, are rarely due to origin from the RAA. There are very few anecdotal reports of similar case presentations. The youngest age group wherein RAA ATs have been characterized was by Friexa X et al., when they had reported a mean age of  $32 \pm 13.6$  years [3]. Sakaguchi et al. had reported ectopic ATs in pediatric age group with a mean of  $8.7 \pm 4.5$  years including a note on RAA AT in a 9-month-old child [4]. Way back in 1992, Walsh et al. had reported RAA ATs in the pediatric age group, however the youngest of them was 7 years of age [5]. In a recent series of 42 patients with appendage ATs from two different centers, 30 patients underwent successful RFA—the youngest of which was 10 years of age. In the same series, 12 (28.6%) patients

underwent minimal invasive surgical appendectomy, the youngest of which was a 5-year-old [6].

RAA atrial tachycardias (ATs) constitute 0.6%–8% of focal ATs, as reported in various case series. These ATs are notoriously incessant in 75% cases and may lead to TCMP in around 50% cases [2–7]. RAA ATs are said to be more common in males and in older children and adults, unlike our case. Precluding the very young age of presentation, our case is similar to the characteristics of RAA ATs described in a few case series: origin of RAA ATs from the base of the RAA, incessant tachycardia, and associated TCMP [8]. Left atrial appendage tachycardias have also shown to have similar clinical characteristics as of RAA ATs [9,10]. Automaticity has been proposed as the mechanism in these ATs originating from the appendages.

In our case, the initial electrophysiology maneuvers included deciphering the mechanism of the tachyarrhythmia and making a convincing diagnosis of focal atrial tachycardia. Reentry mechanism

in the tachycardia was ruled out by a very short bi-atrial activation time, long post-pacing intervals during the overdrive stimuli from the various sites of the tricuspid annulus, the proximal and distal CS electrodes and a centrifugal right atrial activation pattern from the base of the RAA which was confirmed by the 3-D electroanatomical map. In addition, we observed short post-pacing intervals (PPI) at the base of RAA. The short post-pacing intervals, though may be variable in tachycardias of ectopic origin in contrast to reentrant circuits, have shown to relate to the origin of the focal tachycardia [11]. The left atrium was not mapped as the source of the tachycardia from the right atrial appendage was confirmed by the P wave morphology and point-by-point right atrial activation mapping. The P wave morphology has been studied to predict the source of ectopic atrial tachycardias. The negativity in V1 with a slow P wave progression across the chest leads has shown to predict RAA origin of the ectopic with a sensitivity and specificity of 100% and 98% respectively [2,7].

Right atrial appendage is heavily trabeculated with pectinate muscles. Achieving high powers during RFA can be a challenge due to the stagnation of blood flow in the deep recesses of the RAA. At the same time, one should exercise caution in delicately maneuvering the ablation catheter within the RAA, as the risk of perforation remains high. Open-irrigated catheters help in achieving higher powers and at the same time ensuring an effective and durable lesion at such sites [5]. Up-titration of powers, as seen in our case, can help in safe delivery of these lesions.

The 3-D electroanatomical mapping was extremely useful in this case. The child was guarded against radiation exposure by totally utilizing the NavX system for placement of all the catheters, anatomical and activation mapping of the right atrium. The anatomy of the right atrium and the appendage was created using 3-D anatomical mapping. An aneurysm in the RAA was excluded with a transthoracic echocardiography and later with an angiography. RAA aneurysms are known to result in RAA ATs and usually result in recurrences necessitating surgical appendectomies [12–14].

Our patient has been followed for 9 months with no recurrence. Distal source of appendage ATs and associated aneurysms are the factors associated with recurrences, whose rates vary from 20% to 66.7% [12–14]. Incessant ectopic atrial tachycardia accounts for 5% of cases of TCMP in adults and 14% in children [15]. Younger age and persistent nature are recognized as independent risk factors of TCMP. TCMP is usually misdiagnosed in children and usually regarded as dilated cardiomyopathy (DCMP) as a sequel to myocarditis or autoimmune cardiomyopathy. Paying attention to the ECG and awareness of persistent arrhythmias leading to such an entity has to be kept in mind. This is because one can usually expect a near normal recovery of the child's ventricular functions within 1–6 months upon successful treatment of these arrhythmias, in contrast to DCMP. In our case, expectedly, the baby's symptoms of heart failure had improved dramatically after the procedure and the left ventricular functions had returned to near normal within one month of the procedure. Thus, radiofrequency ablation offers cure for such incessant tachyarrhythmias leading to TCMP in children.

#### 4. Conclusion

We have described a rare case of RAA AT leading to TCMP in a 4-year-old child, who had recovered completely from the illness

following a successful radiofrequency ablation. The rarity of RAA ATs in very young children, successful RFA using 3-D electroanatomical mapping and sparing use of fluoroscopy remain the highlights of this case.

#### Declaration of interests

None.

#### Acknowledgement

The authors acknowledge the contributions from our support staff in care of the patient, data compilation, and preparation of figures.

This research did not receive any specific grant from funding agencies in the public, commercial, or not-for-profit sectors.

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