An unusual cause of lone atrial fibrillation in a young female subject due to a rapid-cycling focal atrial trigger

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Introduction

Lone atrial fibrillation (AF) refers to the occurrence of AF in patients with structurally normal heart with normal function.[1] Lone AF is uncommon in children, with an estimated prevalence of 7.5/100,000 patients, though it appears to be more common with increasing age, in male subjects, and in obese children.[1–3] Electrophysiology (EP) studies have been performed in refractory AF in this population; however, there is conflicting information regarding the frequency and location of focal triggers for AF and outcome of ablation.[1,3–6] Here we report on a pediatric patient with refractory AF who had an excellent outcome after catheter ablation of a focal atrial trigger with a location and activation frequency that has never been reported before, and review the available literature on this topic.

Case report

The patient, a 13-year-old nonobese (body mass index: 20.05 kg/m^2) female subject with no significant prior medical history or family history of arrhythmias, was noted at a well child visit to have an irregular heart rhythm by her pediatrician. She was sent for evaluation at a cardiology clinic in an outside hospital, where she was found to be in AF (Figure 1A). She was admitted to the same hospital and she was cardioverted to sinus rhythm after documentation of the absence of intracardiac thrombus by echocardiogram. However, she had recurrence of AF on the same day during observation. At this point she was transferred to our institution, where we started flecainide, and cardioversion was repeated after several doses of this medication, with successful conversion to sinus rhythm. AF again recurred within 12 hours. This series of events was repeated with a trial of sotalol and then a trial of disopyramide, again with similar results: sinus rhythm for 10–12 hours after cardioversion. Her AF would recur in each case in the early morning hours while she was sleeping. Her ventricular rate at the time of recurrence of AF was typically in the 60–70 beats per minute range and the rhythm alternated from an organized atrial tachycardia to AF (Figure 1B and C). The atrial tachycardia, when organized, appeared to have a left atrial focus.

She next underwent an elective EP study and right-heart catheterization to evaluate for other causes of AF, including presence of an accessory pathway, atrioventricular (AV) reentrant tachycardia (AVRT), AV nodal reentrant tachycardia, or inducible atrial tachycardia. She was brought to the EP laboratory in AF and was cardioverted. She remained in sinus rhythm throughout the study, which demonstrated normal hemodynamics. The EP study demonstrated normal AV nodal physiology with no evidence of dual AV nodal physiology, accessory AV connection (manifest or concealed), or inducible tachycardia, including atrial tachycardia or AF, both at baseline and during isoproterenol infusion at 0.015 μG/kg/min. Following the procedure, she was discharged home in sinus rhythm on an amiodarone load. The next morning her parents recorded her heart rhythm using an AliveCor Kardia system (AliveCor Inc, Mountain View, CA), which demonstrated her to be in AF. Another attempt was made to cardiovert her after a load of amiodarone for 1 month, but her AF again occurred early the next morning. Cardiac magnetic resonance imaging was done at this point and did not demonstrate any evidence of atrial fibrosis or scar.

After this recurrence on the amiodarone load, the decision was made to have her undergo repeat EP testing with potential pulmonary vein isolation (or partial pulmonary vein isolation—left pulmonary veins) for AF at our partner adult institution. During this procedure, mapping of her atria while in AF demonstrated an area just proximal to the mouth of her left atrial appendage at the location of the ligament of Marshall with extremely rapid and irregular electrical far-field signals with a cycle length of 46–50 ms

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KEY TEACHING POINTS

- Lone atrial fibrillation is uncommon in children but can be recurrent and refractory to medical management.
- Electrophysiology study in these pediatric population reveal focal triggers or demonstrate atrial fibrillation associated with more common forms of supraventricular tachycardia such as atrioventricular reentrant tachycardia.
- Catheter ablation in this situation appears to be safe, with excellent immediate results.
- A rapidly cycling focal atrial trigger in the region of the ligament of Marshall can present with pediatric lone atrial fibrillation and respond well to catheter ablation.

(Figures 2A–C and 3). Though there was always a rapid signal in this location, the local near-field and the distal atrial signal on the coronary sinus catheter alternated from this similar rapid signal to a more organized and regular atrial tachycardia (Figure 2A–C). Radiofrequency (RF) ablation at this site resulted in prolongation of the cycle length to 80 ms prior to sudden termination of the tachycardia and resumption of sinus rhythm (Figure 2D). This rapid tachycardia was no longer present throughout the rest of the study, including testing with high-dose isoproterenol. She did have a slower organized atrial tachycardia that was mapped to a similar location at the base of the left atrial appendage, which was successfully ablated with RF energy. At the end of the study she had no inducible tachycardia. The following morning, she had an episode of regular atrial tachycardia (same morphology seen in the EP study) that was treated with a single dose of amiodarone, which terminated her tachycardia (Figure 1D). She remained in sinus rhythm without recurrence in the early morning hours and was discharged home. She had a recurrence of this regular atrial tachycardia (cycle length of 320 ms), for which she was symptomatic at home later that day. She was admitted to our institution and started on flecainide, which terminated her tachycardia. She remained on flecainide for 6 months, at which point this medication was discontinued. She has had no recurrence of her tachycardia for the past 5 months. She has had no recorded atrial tachycardia or fibrillation and has recorded her heart rhythm several times with an AliveCor Kardia system (AliveCor Inc, Mountain View, CA).

Discussion

Lone AF is uncommon in children and can be recurrent, as in our patient (recurrence rate: 18%–31% at 3 months).[2,3] Although the underlying trigger for AF in this population is unclear, it has been considered to be multifactorial, with obesity as a recognized risk factor.[2] Familial aggregation of lone AF has also led to identification of an underlying genetic

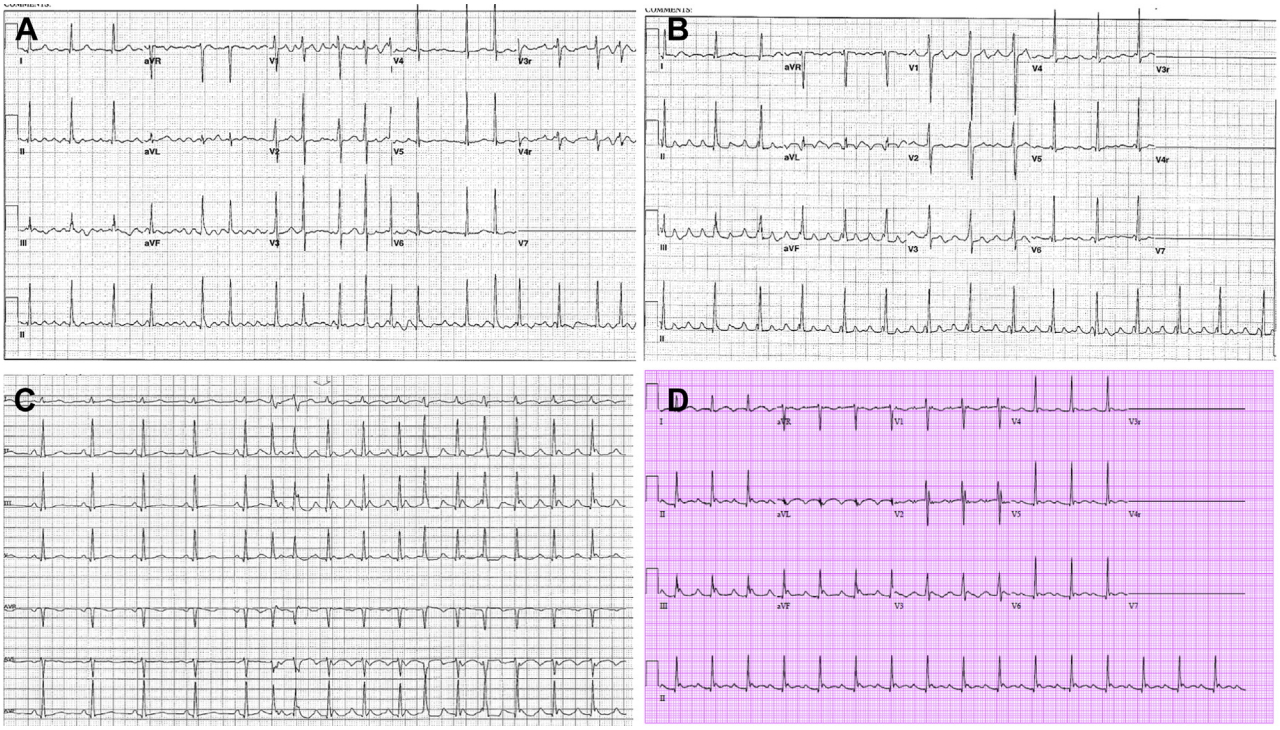

Figure 1 **A:** Electrocardiogram (ECG) on initial presentation, demonstrating disorganized atrial rhythm with variable A-V conduction consistent with atrial fibrillation. **B:** ECG demonstrating organized atrial activity with 2:1 A-V conduction consistent with atrial flutter. **C:** ECG demonstrating initiation of tachycardia with organized atrial rhythm. **D:** ECG demonstrating slower atrial tachycardia with 2:1 A-V conduction after focal radiofrequency ablation.