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Reversible Symptomatic Dilated Cardiomyopathy in Older Children and Young Adolescents Due to Primary Non-Sinus Supraventricular Tachyarrhythmias

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Abstract. Dilated cardiomyopathy (DCM) due to a primary supraventricular tachycardia not originating from the sinus node is not frequently seen in older children or adolescents. However, it is important to recognize this entity as a reversible cause of DCM to avoid costly and inappropriate treatments for these patients. We describe 7 patients who presented with DCM. Five were misdiagnosed as having "sinus" tachycardia secondary to an idiopathic DCM, and 2 were correctly diagnosed as having DCM secondary to an atrial tachycardia. All underwent electrophysiologic treatment of the tachycardia with remission of the DCM.

Key words: Heart failure — Pediatrics — Radiofrequency ablation — Dilated cardiomyopathy — Tachyarrhythmia

Non-sinus supraventricular tachycardia (SVT), defined as a rapid heart rate for age secondary to an abnormal mechanism originating proximal to the bifurcation of the bundle of His, is the most common symptomatic dysrhythmia in childhood and may present at any age [7, 9–11], including the fetus [19, 20]. A reversible form of DCM can occur with chronic arrhythmias and is referred to as "tachycardiomyopathy" [5]. Cessation of the arrhythmia by drug therapy [17, 19], radiofrequency ablation (RFA) of the tachycardia circuit [4, 12–14], or surgical ablation [15] results in the recovery of myocardial function even if the arrhythmia is of long-standing duration. Although SVT-induced DCM is a well-recognized entity in adults with chronic atrial and ventricular tachycardias [1, 5, 13, 15] as well as in infants [8, 9, 20], its occurrence in older children and adolescents is rare [4, 12, 14]. The purpose of this paper is to describe the clinical course of a group of older children and adolescents who presented with DCM secondary to an initially unrecognized non-sinus SVT, reminding the pediatric cardiology community of this reversible cause of DCM.

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Materials and Methods

Patient Selection

In a retrospective review, our combined patients from 1985 to the present who presented to our institutions with tachyarrhythmia and symptomatic DCM were analyzed. All required the prerequisite of absence of previous episodes of tachyarrhythmia and/or structural congenital heart defects. A total of 7 patients, 6 males and 1 female, ages 7–14 years (median 12 years) were identified.

Electrophysiology Study/Radiofrequency Ablation

Initial drug therapy consisted of digoxin, furosemide, and spironolactone in all, and amiodarone and sotalol in 2 patients, respectively (patients **C** and **G**) (Table 1). However, due to persistent clinical symptoms and intractable tachycardia following initial diagnostic evaluation and medical treatment, all patients were referred to a cardiac electrophysiologist for more detailed electrocardiographic analysis. Intracardiac electrophysiologic (EP) studies with RFA were performed in all after obtaining written informed consent. Mapping techniques included intracardiac electrograms in 6, and three-dimensional computer reconstruction of the tachycardia wave dispersion in 1 patient.

Patient Follow-up

After suppressing the SVT with RFA in 5, and with additional antiarrhythmic therapy in 2, 6 patients were followed at regular intervals. At the time of this paper, all patients had received follow-up EGG, exercise stress tests, and serial echocardiograms 3–6 months after initial presentation.

Table 1. Patient characteristics and diagnosis before and after radiofrequency ablation

Patient	Age (years)	HR (bpm)	Diagnosis pre-RFA	Diagnosis post-RFA
A	7	200	AET-induced DCM	AET focus from the crista of the RA.
B	9	234	Viral myocarditis - Sinus tachycardia	Orthodromic AVRT with R septal bypass tract.
C	9	167	Idiopathic DCM - Sinus tachycardia	Multiple atrial muscle macro-reentry circuits: AF from base of RAA (all with distinct cycle lengths). JET. SN dysfunction (prior to RFA)
D	12	167	Viral myocarditis - Sinus tachycardia	AET focus from low antero-septal region of the RA.
E	13	190	Idiopathic DCM - Sinus tachycardia	AET focus from the posterior septum of the RA.
F	13	135	Viral myocarditis - Sinus tachycardia	AET focus from the anterior septum of the RA.
G	14	190	Non-sinus SVT-induced DCM	Atrial muscle macro-reentry circuit from para-SN region.

HR (bpm), heart rate in beats per minute; RFA, radiofrequency ablation; AVRT, atrio-ventricular reciprocating tachycardia; AET, atrial ectopic tachycardia; AF, atrial flutter; RAA, right atrial appendage; CS, coronary sinus; JET, junctional ectopic tachycardia; SN, sinus node; RFA, radiofrequency ablation; DCM, dilated cardiomyopathy; SVT, supraventricular tachycardia.

Left Ventricular Function Assessment

Left ventricular shortening fraction (LVSF) and end-diastolic dimension (LVEDD) were assessed by two-dimensional echocardiography within days prior to and after 3–6 months (median 5 months) following the ablation procedure.

Patient Functional Status

Clinical status was assessed by the New York Heart Association (NYHA) CHF (congenstive heart failure) classification prior to and one month after RFA.

Statistics

Results are reported as median values. The paired Student *t*-test was used to compare measured indices before and after RFA. Significance was defined at $p < 0.05$. The Wilcoxon signed ranks test was used for ordinal data such as NYHA CHF class, with significance defined at $p < 0.05$.

Results

Patient Characteristics

Admission ECG showed narrow QRS tachycardia in all patients, with a heart rate ranging from 135 to 234/min (median 190/min). Two patients presented to our institutions with a diagnosis of primary DCM, 3 presented with a presumed diagnosis of viral myocarditis causing secondary DCM, and 2 were correctly diagnosed as having primary non-sinus SVT with secondary DCM. In 5 patients the tachycardia was believed by the admitting physicians to be sinus in origin (Table 1). The definitive diagnosis of primary non-sinus SVT with secondary DCM was made after careful examination of the surface EGG by the electrophysiologists who identified variable P wave morphology in all 5 patients, 1 of whom had first degree A-V block (Fig. 1a). SVT with absent P waves and atrial ectopic tachycardia were diagnosed in 2 patients, respectively. Based on their clinical presentation, 3 patients were initially classified as NYHA class II, 2 as NYHA class III, and 2 as NYHA class IV. LVSF ranged from 10 to 30% (median 20%) and LVEDD ranged from 4.75 to 7.98 cm (median 5 cm).

Electrophysiologic Studies and Radiofrequency Ablation

EP studies revealed atrioventricular reciprocating tachycardia with septal bypass tract in 1 patient, atrial ectopic tachycardia in 4, and atrial muscle reentry tachycardia in 2, 1 of whom also exhibited junctional ectopic tachycardia and sinus node dysfunction (Table 1). RFA restored sinus rhythm in all but 1 patient who had sinus node dysfunction prior to the RFA procedure (Fig. 1b).

Follow-up of LV Function After RFA

LVSF improved from 10–30% (median 20%) to 31–38% (median 32%, $p = 0.003$), and LVEDD decreased from 4.75–7.98 cm (median 5.7 cm) to 4.3–6.4 cm (median 5 cm, $p = 0.02$) after RFA (Fig. 2). Exercise stress test showed no inducible dysrhythmia in any patient.

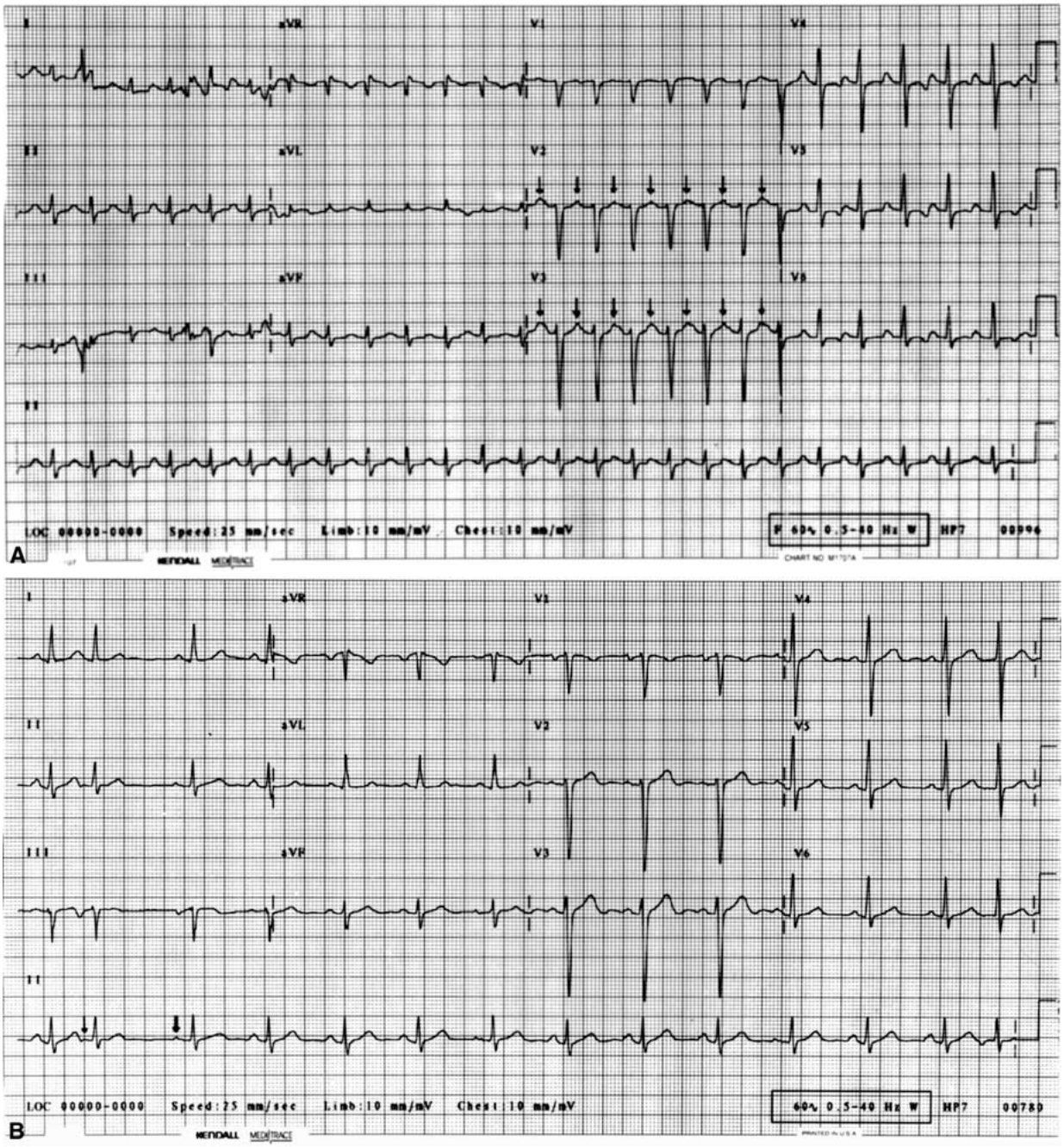

Fig. 1. (**A**) Electrocardiogram on admission. A 12-lead EGG from a 13-year-old boy who presented with CHF. He was misdiagnosed as having viral myocarditis with "sinus tachycardia at a rate of 160/min with first degree atrioventricular block with P waves buried in the preceding QRS." Even though P waves can be seen on the preceding T waves in precordial leads V2 and V3 (*arrows*), they are not consistently seen in lead II. When present, P wave morphology is not distinct, therefore the frontal axis is not clear. These three factors discard the diagnosis of sinus origin of the tachycardia with first degree atrioventricular block in favor of an ill-defined atrial rhythm. (**B**) A 12-lead ECG 3 months later from the same patient after radiofrequency ablation of the non-sinus tachycardia focus. Sinus rhythm is now evident. Lead II shows two normally conducted atrial premature beats with one followed by a normally conducted escape beat (*arrows*), perhaps from the same focus. P waves can be clearly seen now preceding the QRS in all leads.

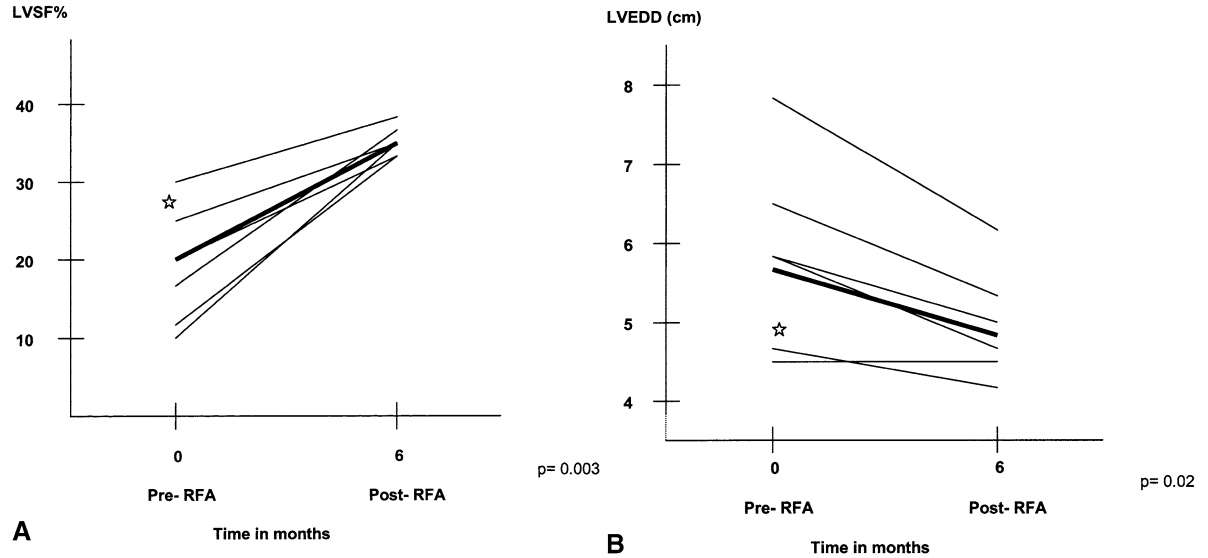

Fig. 2. (**A**) Left ventricular shortening fraction. Among all patients, the LVSF improved significantly after RFA and/or antiarrhythmic medical therapy. Six months after RFA, the left ventricular shortening fraction improved from a median of 16 to 32% ($p = 0.003$). (**B**) Left ventricular end diastolic diameter. Although the LVEDD decreased significantly 6 months after RFA ($p = 0.02$), the LV was still considered to be dilated when compared with normal values for height and weight. The median LVEDD decreased from 5.7 cm to the final median value of 5 cm.

Resolution of Congestive Heart Failure

In 5 of 7 patients, inotropic and antiarrhythmic medications were discontinued after RFA. One patient who was found to have sinus node dysfunction at the time of the EP study remained in junctional/low atrial rhythm with occasional episodes of SVT at a slower rate than on initial presentation (from 190/min before RFA to 100/min after RFA). The intermittent dysrrhythmia, however, was well tolerated and did not hinder this patient's recovery from CHF. He was receiving digoxin, enalapril, amiodarone, and aspirin at the time of his latest follow-up. One patient with atrial ectopic tachycardia required sotalol and digoxin after RFA failed to completely suppress the atrial tachycardia. This patient was in sinus rhythm at the time of follow-up. Prior to correct diagnosis of non-sinus SVT and effective therapy by RFA and antiarrhythmic medication, 2 patients were considered NYHA IV and 2 were considered NYHA III for heart transplantation. Following treatment, all 7 patients became asymptomatic within 1–2 months (NYHA class I).

Discussion

Clinical presentations of non-sinus SVT vary from hydrops and CHF in the newborn [8, 20] to episodes of symptomatic tachycardia or CHF and DCM in children, adolescents [4, 7], and adults [1, 5, 13]. Mechanisms of SVT include reentry and automaticity. In reentrant tachycardia, abnormal electrical impulses that originate within one area of the atria reenter previously depolarized atrial tissue, and at times utilize anatomic para-AV nodal tissue or accessory pathways, creating a macro-reentry circuit within the atrium. An automatic or ectopic SVT results from a rapid and sustained discharge from one or more localized ectopic foci [6]. In our study, 4 patients presented with an atrial ectopic SVT, and 3 with atrial reentrant tachycardias (2 atrial muscle macro-reentry and 1 AV reciprocating tachycardia with an aberrant pathway). One patient presented multiple atrial muscle macro-reentry circuits in addition to a junctional ectopic tachycardia. Due to the unusual nature of this SVT, a viral myocarditis was suspected, although not confirmed with serologic testing (patient **C**) (Table 1). Persistent or incessant tachyarrhythmias not only produce regional or global myocardial contractile abnormalities but can also provoke myocardial damage with fibrosis and adipose infiltration [1–3].

Following suppression of the atrial tachyarrhythmia with antiarrhythmic therapy and/or RFA, cardiac function normalizes. In pediatric [4, 12, 14, 17] and adult studies [1, 5, 13, 15] of atrial dysrrhythmias clinical improvement (assessed by the NYHA CHF classification) and echocardiographic evidence of recovery of LV systolic dysfunction (assessed by the LVSF) occurred after atrial tachycardia supression. In this study, medical therapy with digitalis and

diuretics was initiated, prior to RFA of the atrial tachycardia foci/circuits in all patients, including those initially diagnosed as having viral myocarditis. However, medical therapy is unsuccessful in more than 50% of the patients with atrial ectopic tachycardias (which occurred in 4 of the 7 patients).

Drug therapy is also usually ineffective in modifying the macro-reentry circuit and preventing its recurrence in atrial flutter [16]. In our patient with orthodromic AV reciprocating tachycardia (who received digitalis initially), a more appropriate treatment would have been a beta or a calcium channel blocker. Therefore, it is most likely that only after elucidating the exact tachyarrhytmia mechanism (which was only achieved with the electrophysiologic study in 6 of the 7 patients) an effective antiarrhythmic drug was chosen. The patient in whom the electrophysiology study uncovered multiple atrial muscle macro-reentry circuits and a junctional ectopic tachycardia (patient **C**) (Table 1), RFA was ineffective in terminating the tachycardia. However, by modifying the tachycardia circuits, RFA allowed the antiarrhythmic medications (digoxin and amiodarone) to achieve complete control of the rapid heart rate. In our study, cardiac function normalized in all patients who we were able to be followed up (6 of 7).

In spite of significant regression in the LV size (measured by the LVEDD) 6 months after cessation/control of the atrial tachycardia, the LVEDD continued to exceed the upper limits of normal for the patients' height and weight in all cases. This occurs because sustained, chronic tachycardia diminishes the number of microtubules in the cardiac myocyte, which contributes to myocardial contractile dysfunction [18]. Due to depletion of high-energy phosphates, LV function requires a minimum of 1 month after cessation of the tachycardia to improve. Recovery of ventricular function is typically followed by a slower recuperation of LV size that peaks after 6–8 months [5]. This delay in LV size normalization after resolution of the atrial tachycardia has been explained from animal studies by demonstrating collagen decrease in the extracellular matrix in tachycardiomyopathy specimens, causing less adhesiveness of the basal membrane to the myocytes and persistent intracellular spacing [18]. Therefore, while cardiac contractility improves rapidly (shown by resolution of clinical symptoms of CHF and by early enhancement in LVSF) after cessation of the tachycardia, correction of LV dilatation (measured by the LVEDD) is a slower phenomenon, based on anatomical and histologic remodeling of the heart.

Sinus tachycardia, secondary to inflammatory processes of the myocardium leading to DCM and CHF, is more common among older children and adolescents than DCM secondary to tachyarrhythmia. Consequently, these patients can be easily misdiagnosed, potentially leading to inappropriate and costly treatments, including unnecessary listing for heart transplant. Therefore, the older child or adolescent who presents with DCM and persistent tachycardia should be thoroughly evaluated for a primary atrial tachyarrhythmia. As an example, one of our patients was initially diagnosed as having sinus tachycardia with first degree A-V block, which would be very unusual because physiologic sinus tachycardia is often associated with enhanced A-V conduction (unless A-V conduction abnormalities exist). Therefore, in the setting of a symptomatic patient with DCM, the ECG appearance of a sinus tachycardia with first degree A-V block may indicate that the atrial tachycardia is non-sinus. Consequently, ECG analysis of a patient with tachycardia and DCM requires careful evaluation, looking for absence of P waves, examining P wave morphology when present, and paying close attention to the duration of the PR interval. This can lead to a correct diagnosis and will permit proper initial therapy for this reversible cause of heart failure.

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