

Idiopathic Left Ventricular Tachycardia in Children

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Background: Idiopathic left ventricular tachycardia (ILVT) is a rare but well-recognized clinical entity. The clinical characteristics and prognosis of this form of ventricular tachycardia (VT) in Thai children is not known.

Objective: To define clinical presentations, drug therapies, roles of radiofrequency (RF) catheter ablation, and the short-term outcome of these children in Thailand.

Patients and Method: From April 1999 to June 2007, 10 patients were diagnosed as ILVT by specific electrocardiographic features and therapeutic response. All patients had a structurally normal heart. Data were collected retrospectively. Baseline clinical information, 12-lead electrocardiography (ECG) during VT, responses to drug therapy, results of RF catheter ablation therapy, and outcome were determined.

Results: Median age at presentation was 9.5 years (range, 3.8 to 14.0 years). Three patients (30%) were male. Eight patients (80%) were diagnosed as supraventricular tachycardia (SVT) before ILVT diagnosis. Median duration from SVT diagnosis to the correct diagnosis was 1.5 years (range, 0 to 6.0 years). Palpitation and chest pain were usual clinical manifestations while congestive heart failure was the presentation in one due to incessant tachycardia. Two patients had recurrent VT episodes during acute febrile illnesses. The majority of patients responded to intravenous verapamil. RF catheter ablation was performed in 3 patients with recurrence of the VT in one.

Conclusion: Prompt recognition of the ILVT especially in the emergency department is very important. Verapamil is effective for acute termination as well as prevention of VT recurrence. When VT is refractory to medical therapy, RF catheter ablation is safe and effective. The short-term prognosis was good.

Keywords: Ventricular tachycardia, Pediatrics, Children, Arrhythmia

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Ventricular tachycardia (VT) in children with apparently normal heart is uncommon. Idiopathic left ventricular tachycardia (ILVT) is a distinct subgroup

of idiopathic VT that may be confused with either typical VT or supraventricular tachycardia (SVT). In 1979, Zipes et al⁽¹⁾ reported VT in 3 young patients who were characterized by QRS width of 120 to 140 milliseconds (msec), right bundle branch block morphology, and left-axis deviation. These patients had no structural heart disease. Belhassen et al⁽²⁾ observed that this tachycardia can be terminated by the calcium

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channel blocker, verapamil. Despite being VT, the prognosis for these patients is generally excellent. The authors summarized their experience in the diagnostic clues, management, and prognosis during short-term follow-up of this rare form of VT in children.

Material and Method

Inclusion criteria

This is a retrospective study from April 1999 to June 2007. A total of 10 patients at King Chulalongkorn Memorial Hospital with the diagnosis of ILVT were identified. Data were gathered from medical records and the database of the division of cardiology, department of pediatrics and the cardiac catheterization laboratory. The diagnosis was made by using the following criteria: 1) electrocardiographically documented sustained monomorphic VT originating from the left ventricle (right bundle branch block morphology and left-axis deviation) (Fig. 1); 2) no apparent structural heart disease; and 3) termination of VT by verapamil or diltiazem (Fig. 2, middle). The absence of organic heart disease was diagnosed on the basis of 1) normal findings on cardiac examination; 2) normal findings on the rest electrocardiography (ECG) and chest X-ray; and 3) normal findings on the echocardiogram. Demographic and clinical data including baseline clinical information, 12-lead ECG during VT, responses to drug therapy, results of radiofrequency (RF) catheter ablation, and outcome were collected in June 2007.

Follow-up

Patients were followed up from the date of presentation to the last day of the present study. The majority of patients underwent regular outpatient visit at King Chulalongkorn Memorial Hospital or the referring cardiac centers. Data were obtained by phone in a few patients who experienced no tachycardia recurrence and who were lost to follow-up.

Statistical analysis

Quantitative data were expressed as mean \pm SD or as median, range, and percentage as appropriate. Statistical analysis was performed using a commercial statistic software package (SPSS Inc., Chicago, IL, USA).

Results

Ten patients were diagnosed with ILVT from 1999 - 2007. The clinical characteristics and follow-up data of all patients are summarized in Table 1. The age at presentation ranged from 3.8 to 14.0 years (median, 9.5 years) and 30% were male. Eight patients (80%)

were diagnosed as SVT before ILVT diagnosis. Median duration from SVT diagnosis to the correct diagnosis was 1.5 years (range, 0 to 6.0 years).

Symptoms at presentation

Palpitation and chest pain were the usual clinical presentations. Congestive heart failure was found in 1 patient (case No. 5) in whom the VT resisted to intravenous lidocaine therapy. After control of the VT with cardioversion followed by RF catheter ablation, left ventricular ejection fraction in this patient improved from 23% to 74%. One patient (case No. 4) presented with a history of vomiting without palpitation during the tachycardia. In 2 patients (20%), the VT episodes were repeatedly triggered by acute febrile illnesses.

Electrophysiologic characteristics

VT rate varied from 120-200 beats per minute (bpm). Relatively narrow QRS complex were demonstrated in the majority of patients. Mean QRS width was 119.6 ± 18.8 msec (range, 80 to 147 msec). QRS configuration during VT was right bundle branch block morphology with left-axis deviation (-60 to -120 degree) in all patients (Fig. 1).

Treatments (Table 2)

Six patients received intravenous adenosine as the first line acute management because of the initial diagnosis of SVT. Adenosine failed to terminate the tachycardia in all patients (Fig. 2, top). Intravenous amiodarone was the second line acute management in 4 patients (case No. 2, 4, 8, 10) in whom the VT slowed down and was finally terminated in 3 patients (duration from administration to effectiveness ranged 1-48 hours). In 1 patient (case No. 2), amiodarone could not terminate the VT and the patient was electrically cardioverted. Intravenous calcium channel blocker (verapamil, or diltiazem) consistently terminated tachycardia in 7 patients (17 episodes) (Fig. 2, middle), typically by slowing the VT rate to less than sinus rate resulting in sinus impulse capturing the ventricle via the atrioventricular (AV) node. Normal QRS axis and narrow QRS complex was documented in all patients after ILVT termination. The nonspecific transient inferolateral T-wave inversion was found in at least 4 of the presented patients. Cardioversions were needed in 4 patients (case No. 1, 2, 4, 5) due to unresponsiveness to medication and/or heart failure. Three episodes were successful. The patient in whom cardioversion (2.5 Joules/kg) failed was given intravenous amiodarone and the VT was under control 48 hours later. Intravenous

Table 1. The demographic data and clinical characteristics of the 10 patients with ILVT

No.	Sex	Age at first presentation (yr)	Age at Dx (yr)	Presentation	Precipitating causes	Previous SVT diagnosis	Response to acute treatment	Long-term treatment	RFA and results
1	F	3.8	6.8	Palpitation	Unknown	Yes	Diltiazem, cardioversion	Verapamil	Not done
2	F	5.0	11.0	Palpitation	Unknown	Yes	Cardioversion	Amiodarone (prior to RFA)	Yes, success
3	F	5.3	7.0	Palpitation, chest pain	Fever	Yes	Verapamil	Verapamil	Not done
4	F	5.4	6.6	Vomit	Unknown	Yes	Amiodarone	Verapamil	Not done
5	M	8.0	13.3	Palpitation, chest pain, CHF	Unknown	Yes	Cardioversion verapamil	Verapamil (after VT recurrence)	Yes, recurrence
6	F	11.0	12.3	Palpitation, chest pain, fatigue	Fever	Yes	Verapamil	Verapamil (prior to RFA)	Yes, success
7	M	12.0	12.0	Palpitation, dizziness	Unknown	Yes	Verapamil	Verapamil	Not done
8	F	12.9	14.9	Palpitation	Unknown	Yes	Amiodarone-diltiazem	Atenolol	Not done
9	F	13.2	13.6	Palpitation	Unknown	No	Verapamil	Verapamil	Not done
10	M	14.0	14.8	Palpitation, chest pain	Unknown	No	Amiodarone	Atenolol	Not done

CHF = congestive heart failure; Dx = diagnosis; ILVT = idiopathic left ventricular tachycardia; RFA = radiofrequency catheter ablation; SVT = supraventricular tachycardia

Table 2. Acute treatments and responsiveness in 10 patients with ILVT

Medications/ cardioversion	Treatment episodes	Number of subjects	Responded episodes	Time to sinus rhythm
Adenosine	9	6	0 (0%)	-
Amiodarone	4	4	3 (75%)	1-48 hours
Verapamil	13	5	13 (100%)	<15 minutes
Diltiazem	4	2	4 (100%)	<15 minutes
Digoxin	2	1	0 (0%)	-
Lidocaine	2	2	0 (0%)	-
Cardioversion	4	4	3 (75%)	Immediate

digoxin could not terminate the tachycardia in 1 patient in whom it was given.

Verapamil was the first line drug therapy for long-term treatment in 6 patients. Dosage of verapamil in the presented patients varied from 1.0 to 8.5 mg/kg/day (mean, 4.5 ± 2.5 mg/kg/day). Atenolol was prescribed in 2 patients (20%).

One patient (case No. 8) had goiter and hyperthyroidism after treatment with oral amiodarone from another hospital for 6 months. Hyperthyroidism

was controlled with methimazole. Mobitz type I second degree AV block was seen in 1 patient who was on verapamil (8 mg/kg/day). The AV block disappeared after the dose was decreased.

Nonpharmacological treatment

Electrophysiologic study (EPS) and RF catheter ablation were performed in 3 patients (case No. 2, 5, 6). Incessant tachycardia with decreased ejection fraction was the indication in case No. 5

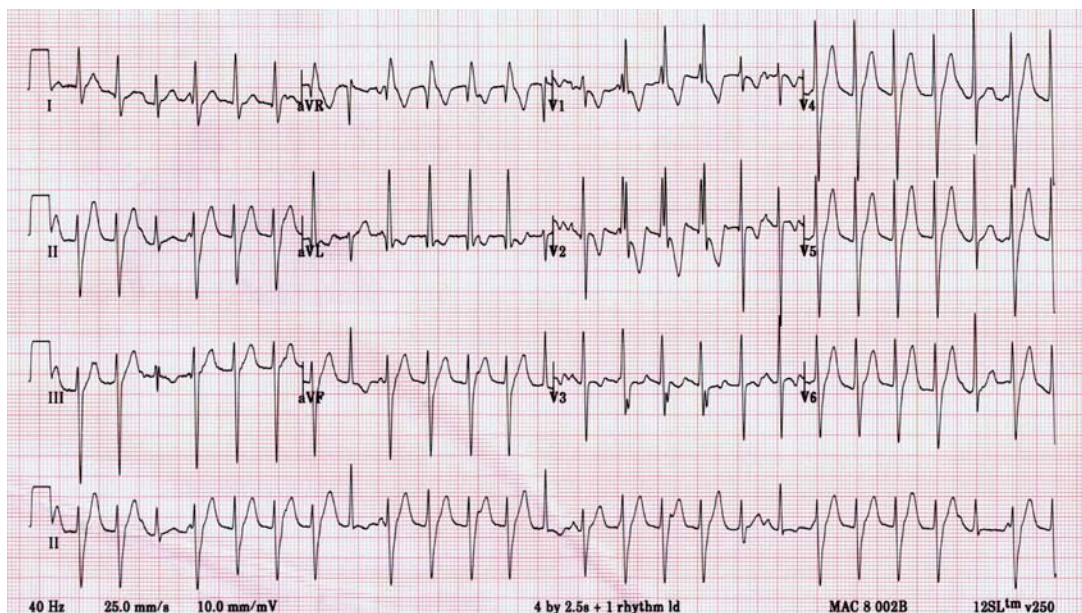


Fig. 1 A 7 year-old girl (case No. 3) presented with palpitation and chest pain during acute febrile illness. 12-lead ECG during tachycardia revealed wide QRS complex tachycardia, QRS width 120 msec, right bundle branch block morphology, and left-axis deviation. Capture beats indicating atrioventricular dissociation also presented

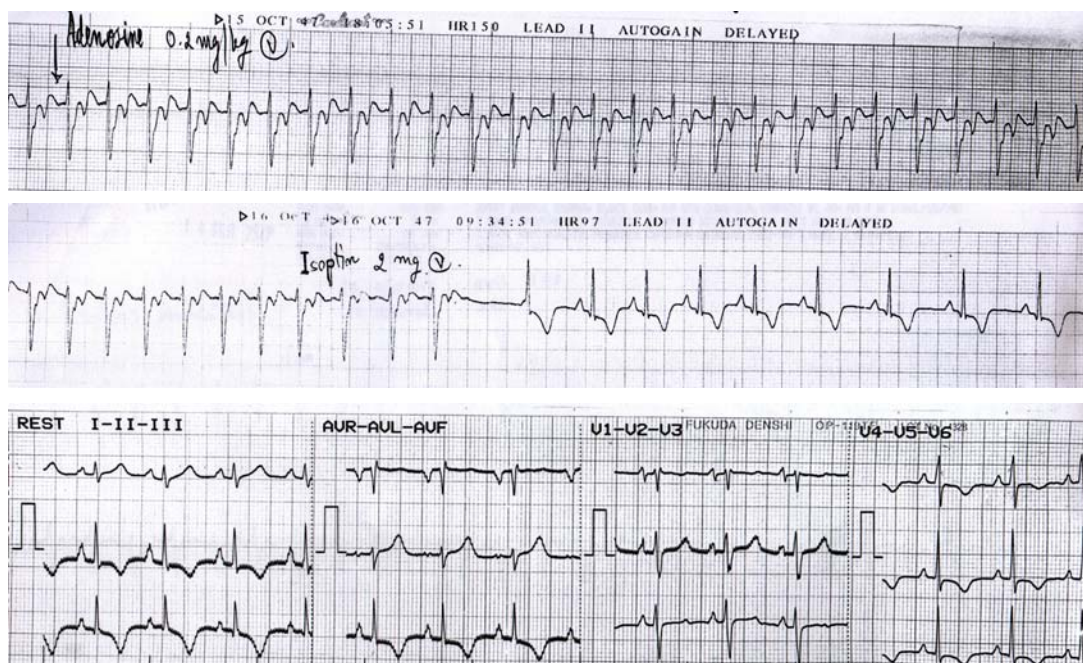


Fig. 2 Electrocardiograms showing ventricular tachycardia in case No. 9. Top, After 0.2 mg/kg of intravenous adenosine, ventricular tachycardia could not be terminated. Middle, After 0.05 mg/kg of intravenous verapamil, VT slowed down and was terminated. Bottom, Sinus rhythm after VT conversion revealed the usual T wave inversion in II, III, aVF, and V₄₋₆

while paroxysmal symptomatic tachycardia was the indication in the other 2 patients⁽³⁾. Acute success was found in all patients. One patient (case No. 5) had recurrent VT 1 month after the procedure which was later controlled by oral verapamil.

Follow-up

Follow-up ranged from 1.3 to 8.1 years (median, 4.5 years). Two patients (40%) who received oral verapamil had recurrent VT. Tachycardia recurrence was found in 1 patient (50%) in oral atenolol group. EPS and RF catheter ablation were an alternative management for these patients.

Discussion

Ventricular tachycardia in the presence of an apparently normal heart (idiopathic VT) is well described in adults but rarely in children. These arrhythmias are usually monomorphic ventricular tachycardias originating from the left ventricle or right ventricular outflow tract. Ventricular tachycardia in the presence of a normal heart is different from VT in patients with structural heart disease in the etiology, clinical presentation, diagnosis, management and most importantly, the prognosis. Published pediatric studies on idiopathic VT have shown a favorable prognosis for the children affected⁽⁴⁻¹¹⁾. ILVT has specific electrocardiographic features and therapeutic options. Most ILVT are verapamil-sensitive intrafascicular tachycardia. Because of its rarity, these arrhythmias are not well known to occur in children and have not been previously described in Thailand.

Fifty percent of our patients presented in the first decade. The youngest patient was 3.8 years old at presentation. Infancy presentation was not found in the present studies as opposed to the previous studies^(11,12). Because of the relatively narrow QRS complex, 80% of the presented patients were initially diagnosed and treated as SVT. Adenosine was often used as the first line medication without termination of the tachycardia (usually, there was no effect on the tachycardia at all). In many instances, this was the first clue that the diagnosis of SVT might have been incorrect. Careful evaluation of the 12-lead ECG during the clinical tachycardia is mandatory in differentiating SVT from ILVT. Wide QRS complex tachycardia with right bundle branch block morphology and left-axis deviation is characteristic for this form of VT. The relatively narrow QRS complex (QRS width 80-147 msec in our study) is the result of intrafascicular re-entry and the relatively small myocardial mass in pediatric age group. The QRS width

is, however, wider than during normal sinus rhythm. However, the QRS width can look narrow in certain leads, such as lead I and lead aVL (Fig. 1). When present, atrioventricular dissociation and/or fusion and capture beats (Fig. 1) differentiate ILVT from SVT as well.

Symptoms of ILVT were usually benign and were not different from the SVT symptoms. The majority of patients had palpitation and/or chest pain. Congestive heart failure and/or syncope were rare, most likely due to the normal ventricular function and the short duration of the VT prior to the patients' seeking medical attention. Generally, ILVT in children and adults have good prognosis^(4-11,13-16). Only one of the presented patients had congestive heart failure with reduced left ventricular systolic function, most likely due to tachycardia-induced cardiomyopathy. The ventricular function improved after successful RF catheter ablation of the VT which is a reversible cause of systolic heart failure⁽¹⁷⁾.

All of the presented patients with sustained VT required medical treatment. Calcium channel blocker sensitivity was characteristic of this disease as previously described⁽²⁾. The dramatic response to calcium channel blocker can again lead to misdiagnosis of SVT. Amiodarone was used in a few patients in whom the diagnosis of ILVT was not recognized, or in whom the Pediatric Advanced Life Support tachycardia algorithm was followed (both in patients who were diagnosed as SVT as well as VT)⁽¹⁸⁾. In the authors' experience, the success of amiodarone administration in terminating ILVT was less than verapamil and long treatment duration might be needed before tachycardia termination. After ILVT termination, the usual non-specific transient inferolateral T-wave inversion was found in the presented patients which did not require any treatment (Fig. 2, bottom). The T wave abnormalities generally returned to normal in a period of weeks to months.

RF catheter ablation can also be an option for acute treatment in patients who do not respond to the medical treatment or cardioversion. In the authors' experience, this method would be needed only rarely since most (if not all) patients can be managed acutely by medical means.

In the authors' experience, verapamil and atenolol were safe and effective for long-term ILVT control in children. Patients who had recurrence despite drug therapy, patients with severe symptoms, or patients who do not want to take medication are candidates for EPS and RF catheter ablation. Because of the report of spontaneous remission up to 50%

in children with ILVT^(6,9,10), the authors have not advocated EPS and RF catheter ablation as the first line treatment in all of the presented patients. A longer follow-up is needed for the long-term prognosis of the presented patients who are currently symptom-free on medication.

Conclusion

Prompt recognition of the ILVT, especially in the emergency department, is important for proper management of this rare arrhythmia in children. When diagnosed and the patient is stable, verapamil is effective for both the acute termination as well as prevention of VT recurrence. Direct current cardioversion is reserved for patients with heart failure or hemodynamic instability. RF catheter ablation is safe and effective for long-term management. Good prognosis can be expected for children with ILVT.

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ภาวะหัวใจเต้นเร็วโดยมีจุดกำเนิดจากห้องล่างซ้ายชนิดไม่ทราบสาเหตุในเด็ก

มนนิภา สื่อเสาวลักษณ์, อภิชัย คงพัฒนโยธิน, บัญชา ศันสนียวิทย์กุล, ทศพร ศิริโสภิตกุล, วรการ พรหมพันธุ์, เยาวลักษณ์ จรรย์พงษ์ไพบูลย์, สุรพันธ์ สิทธิสุข

ภูมิหลัง: ภาวะหัวใจเต้นเร็วโดยมีจุดกำเนิดจากห้องล่างซ้ายชนิดไม่ทราบสาเหตุพบได้น้อยแต่มีลักษณะเฉพาะของโรคในประเทศไทยยังไม่มีการศึกษาเกี่ยวกับการดำเนินโรคและการพยากรณ์โรคของผู้ป่วยเด็กกลุ่มนี้

วัตถุประสงค์: เพื่อประเมินลักษณะทางคลินิก การรักษาทางยา และผลของการรักษาด้วยการจี้จุดกำเนิดภาวะหัวใจเต้นเร็วด้วยคลื่นวิทยุผ่านทางสวนหัวใจ รวมถึงการพยากรณ์โรคระยะสั้นในผู้ป่วยเด็กเหล่านี้ในประเทศไทย

วัสดุและวิธีการ: ทำการศึกษาย้อนหลังในผู้ป่วยที่ได้รับการวินิจฉัยเป็นภาวะหัวใจเต้นเร็วโดยมีจุดกำเนิดจากหัวใจห้องล่างซ้ายชนิดไม่ทราบสาเหตุตั้งแต่เมษายน พ.ศ. 2542 ถึง มิถุนายน พ.ศ. 2550 การวินิจฉัยโดยอาศัยลักษณะเฉพาะจากคลื่นไฟฟ้าหัวใจและการตอบสนองต่อการรักษา ข้อมูลประกอบด้วยลักษณะทางคลินิก คลื่นไฟฟ้าหัวใจ 12 ช่องขณะมีอาการ การตอบสนองต่อการรักษาทางยา และผลของการรักษาด้วยการจี้จุดกำเนิดภาวะหัวใจเต้นเร็วด้วยคลื่นวิทยุ ผ่านทางการสวนหัวใจ

ผลการศึกษา: ผู้ป่วย 10 ราย อายุที่แสดงอาการเริ่มตั้งแต่ 3.8 ปีถึง 14.0 ปี (ค่ามัธยฐาน 9.5 ปี) เป็นผู้ป่วยชาย 3 ราย (30%) ผู้ป่วย 8 ราย (80%) ได้รับการวินิจฉัยเป็นภาวะหัวใจเต้นเร็วโดยมีจุดกำเนิดเหนือต่อหัวใจห้องล่างก่อนที่จะได้รับการวินิจฉัยเป็นภาวะหัวใจเต้นเร็วโดยมีจุดกำเนิดจากหัวใจห้องล่างซ้ายภายในระยะเวลา 0-6.0 ปี (ค่ามัธยฐาน 1.5 ปี) อาการใจสั่นและเจ็บหน้าอกเป็นอาการส่วนใหญ่ที่นำผู้ป่วยมาพบแพทย์ พบภาวะหัวใจล้มเหลวในผู้ป่วย 1 ราย (10%) ซึ่งเกิดจากภาวะหัวใจเต้นเร็วต่อเนื่องเป็นเวลานาน ผู้ป่วย 2 รายมีอาการขณะมีไข้ ผู้ป่วยส่วนใหญ่ตอบสนองต่อการรักษาด้วยยา verapamil ทางหลอดเลือด ผู้ป่วย 3 รายได้รับการรักษาด้วยการจี้จุดกำเนิดภาวะหัวใจเต้นเร็วด้วยคลื่นวิทยุผ่านทางสวนหัวใจ ผู้ป่วย 1 รายพบมีอาการกลับเป็นซ้ำ

สรุป: การวินิจฉัยภาวะหัวใจเต้นเร็วโดยมีจุดกำเนิดจากหัวใจห้องล่างซ้ายชนิดไม่ทราบสาเหตุมีความสำคัญ โดยเฉพาะอย่างยิ่งในภาวะฉุกเฉิน การรักษาด้วยยา verapamil ได้ผลดีทั้งในภาวะเฉียบพลันและการป้องกัน การกลับเป็นซ้ำในระยะยาว กรณีที่ดื้อต่อการรักษาทางยา การรักษาด้วยการจี้จุดกำเนิดภาวะหัวใจเต้นเร็วด้วยคลื่นวิทยุผ่านทางสวนหัวใจมีความปลอดภัยและได้ผลดี การพยากรณ์โรคในระยะสั้นดี
