

Idiopathic Left Ventricular Tachycardia (ILVT) in Children, Co-Incident with Acute Rheumatic Fever

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ABSTRACT

Idiopathic Left Ventricular Tachycardia (ILVT) is a subtype of IVT characterized by right bundle branch block (RBBB) and left axis deviation (LAD) or right axis deviation (RAD) on an electrocardiogram. Idiopathic LVT predominantly occurs in young males with no underlying structural heart disease, metabolic disorders, or genetic abnormalities. However in this case we report a case of 13 years old male patient who has ventricular tachycardia coincident with acute rheumatic fever that was refractory to common therapies such as amiodarone and cardioversion, but resolved after the insertion of a temporary pacemaker and catheter ablation.

Keywords: Idiopathic left ventricular tachycardia, pediatric arrhythmia, catheter ablation, rheumatic fever.

INTRODUCTION

Ventricular tachycardia (VT) occurs in approximately 1 in 100,000 children, with a prevalence of 2 to 8 per 100,000 in school-aged children. Idiopathic ventricular tachycardia (IVT) is the most common VT in healthy children, includes the subtype Idiopathic left ventricular tachycardia (ILVT). Idiopathic left ventricular tachycardia characterized by right bundle

branch block (RBBB) and left axis deviation (LAD), right axis deviation (RAD) or undetermined on electrocardiography (ECG) examination, primarily affects young males without structural heart disease, metabolic disorders, or genetic abnormalities. [2, 3, 4] Idiopathic left ventricular tachycardia is a type of monomorphic tachycardia with various underlying mechanism, including reentry, triggered activity, and automaticity.⁷ One variety of ILVT is fascicular ventricular tachycardia (fascicular VT), which is further classified into three types:[3]

1. **Left Posterior Fascicular VT (LPPFT):** Characterized by RBBB and superior axis deviation.
2. **Left Anterior Fascicular VT (LAFVT):** Characterized by RBBB and RAD.
3. **Upper Septal Fascicular VT (rare):** Typically presents with atypical morphology, usually showing RBBB but may resemble left bundle branch block. Cases with narrow QRS complexes and a normal axis have also been reported.

The most common type of ILVT is left posterior, typically characterized by a narrow QRS complex, which often leads to misdiagnosis as Supraventricular Tachycardia (SVT), approximately 13.5% of VT cases were misdiagnosed as SVT in the emergency department. Standard

treatments such as adenosine, amiodarone, or cardioversion often failed in these patients. Calcium channel blockers (CCBs), including for infants, can be effective in such cases. Ventricular tachycardia should be aggressively treated, with management options including CCBs or radiofrequency catheter ablation, due to the risks of death and tachycardia-induced cardiomyopathy. [4, 5, 6]

Acute Rheumatic Fever (ARF) is a significant inflammatory disease often triggered by a streptococcal throat infection, predominantly in children aged 5–14 years. Diagnosis was established based on Jones criteria. Major criteria include migrant polyarthritis, carditis, Sydenham's chorea, erythema marginatum and subcutaneous nodules. Meanwhile, minor criteria consist of arthralgia, fever, increased erythrocyte sedimentation rate (ESR), C-reactive protein, leucocytosis, and prolongation of the PR interval. The diagnosis of ARF requires either 2 major criteria or 1 major and 2 minor criteria, along with evidence of streptococcal infection (positive throat culture, rapid antigen test, or elevated ASTO titer). Children with ARF, particularly those with recurrent streptococcal infections, face an increased risk of ventricular tachycardia (VT), a rare but serious complication. Studies show VT is significantly more common in children with ARF, highlighting the importance of vigilant cardiac monitoring in these patients. [1]

The occurrence of ILVT concomitant with structural heart abnormalities is very rare. Whether these two conditions are related or coincidentally occur together remains a dilemma. However, this can be clarified after catheter ablation, to determine whether cardiac function improves.

CASE REPORT

A 13 years old male child has chief complained heart pounding since 2 weeks before admission. Three weeks before admission, the patient experienced a high fever that initially subsided with medication

but subsequently recurred. Two weeks before admission, the patient reported sensation of pounding in the chest, both at rest and during activities, along with chest pain and shortness of breath. The shortness of breath was particularly noticeable during episodes of chest pounding.

The patient was referred from other hospital with diagnosed unstable supraventricular tachycardia due to suspected myocarditis, rheumatic heart disease, dilated cardiomyopathy, cardiogenic shock, acute pulmonary edema, bilateral pleural effusion post-taping, acute kidney injury, hypocalcemia, hypokalemia, and metabolic acidosis. Echocardiography revealed decreased left ventricular systolic function with an EF of 39%, eccentric hypertrophy, global hypokinesia, mild to moderate mitral regurgitation, moderate tricuspid regurgitation, and intermediate probability of pulmonary hypertension. The patient had received various medications including ceftriaxone, furosemide, paracetamol, prednisone, benzathine penicillin, amiodarone infusion, calcium, potassium and synchronized cardioversion. He also experienced cardiac arrest while being transported, but returned to spontaneous circulation (ROSC) before arrived the emergency department of Dr. M. Djamil Hospital.

Physical examination revealed the heart rate was 156x/ minute, weak pulse, respiratory rate was 28x/minute, BP was 115/70 mmHg. The beats were regularly substantial in all the four limbs. Heart examination showed ictus was visible, thrill (+), ictus cordis was palpable at 1 finger of lateral left midclavicular line at 6th intercostal space, faster heartbeat, 1st and 2nd heart sounds difficult to assess, systolic murmur gr 3/6, punctum maximum in apex.

The patient's ECG (Figure 1) shows a rate of 158x/minute, LAD, absence of P wave, RBBB, right ventricular hypertrophy, It was suitable with Idiopathic Left Ventricular Tachycardia (ILVT).

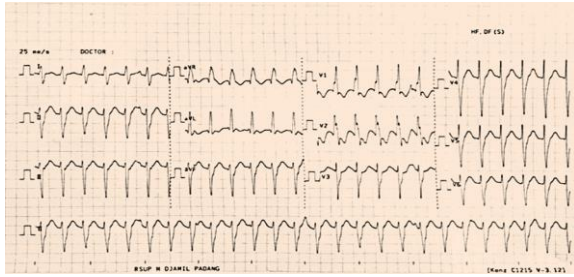


Figure 1 : ECG demonstrates VT, ventricular rate 158x/min, QRS duration 100 ms, RBBB, Left axis deviation, right ventricular hypertrophy.

Chest x-ray examination (Figure 2) on this patient shows Cardiomegaly, CTR 57%

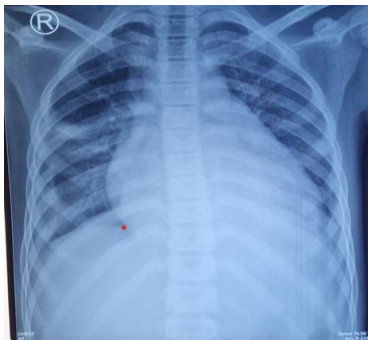


Figure 2 : X-ray chest indicates cardiomegaly.

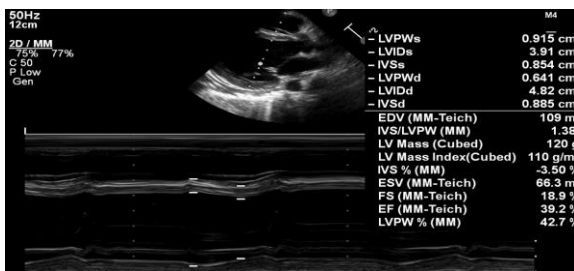


Figure 3 : Echocardiography post catheter ablation, ejection fraction 39,2%

The patient was administered antiarrhythmic therapy with amiodarone infusion for 3 days, bisoprolol and captopril for heart failure therapy, however, the rhythm did not revert to sinus rhythm, and instead, the blood pressure decreased and the patient became unstable. Amiodarone was discontinued, and a dobutamine infusion was initiated. A temporary pacemaker was implanted with a single drive (200 bpm) to control the rhythm. The patient's rhythm subsequently converted to sinus rhythm with a heart rate of 75-100 bpm and the patient no longer required inotropic support.

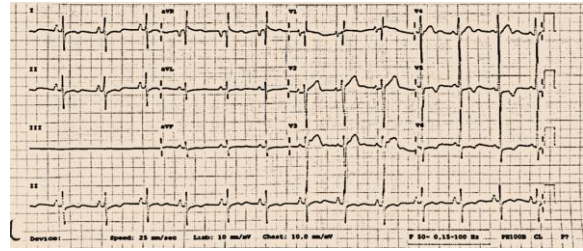


Figure 4. ECG post cardioversion with TPM HR 75x/mnt

The patient experienced a recurrence of ventricular tachycardia after 3 months, with unstable hemodynamics. The patient was unable to tolerate antiarrhythmic therapy with amiodarone or diltiazem due to unstable hemodynamics, verapamil wasn't available in our hospital, During this period, synchronized cardioversion was performed three times, but the rhythm did not revert to sinus rhythm. A temporary pacemaker was then implanted to convert the heart rhythm. The patient was subsequently referred to a larger center for electrophysiological study and catheter ablation.

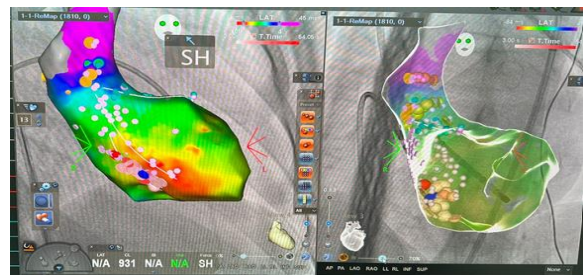


Figure 5. EP study : Suggestive re entry side from distal posterior left fascicle

The results of the electrophysiological study showed an ectopic focus originating from the distal posterior left fascicle. Based on these findings, catheter ablation was performed on the patient.

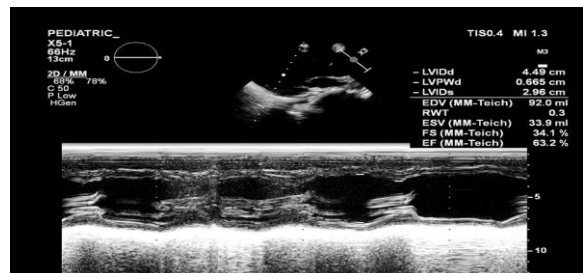


Figure 6. Echocardiography post catheter ablation, ejection fraction 63,2%

DISCUSSION

The patient presented ventricular tachycardia with a narrow QRS complex, initially suspected to be supraventricular tachycardia (SVT), with a history of refractory cardioversion. Subsequently, the child was administered an amiodarone infusion due to the unavailability of adenosine in Indonesia. Despite the administration of an amiodarone infusion, the ventricular tachycardia remained refractory. On the third day, the patient's blood pressure dropped and was hemodynamically unstable. Echocardiography examination showed a decrease in ejection fraction to 39%, and the result of USCOM examination showed low preload, low inotropy, and normal after load, the patient was assessed with cardiogenic shock and received a dobutamine infusion. Then the patient was consulted to the heart electrophysiology division, based on the ECG findings and history of refractoriness to antiarrhythmic drugs and cardioversion, the patient was diagnosed with ILVT concomitant with ARF. The patient was implanted TPM. Cardioversion was performed using TPM, with overdrive pacing several times at a frequency of 200 bpm. The rhythm returned to sinus with the rate 80-90 bpm. The patient was advised to be referred to a more advanced healthcare facility for electrophysiology studies and catheter ablation; however, the family declined due to financial reasons and was discharged with bisoprolol, therapy for DRA continued in outpatient clinic. The ECG examination in our hospital showed ventricular tachycardia (VT) RBBB and LAD, consistent with ILVT. Idiopathic left ventricular tachycardia typically occurs in young male adults, with the most common clinical presentations being paroxysmal episodes of palpitations, dizziness, and less frequently, syncope. The electrocardiographic pattern of ILVT varies depending on the site of origin of the tachycardia. Posterior ILVT accounts for 90-95% of cases and is characterized by an RBBB morphology and LAD, it is often

misdiagnosed as SVT and frequently resistant to treatment with adenosine, amiodarone, and direct current cardioversion, which can lead to ventricular dysfunction.[4, 7, 8, 9]

Unstable ILVT cases which is unresponsive to therapy, electrical cardioversion can be performed, though it may become ineffective with repeated use, as seen in this patient. For severe symptoms unresponsive to pharmacological treatment, catheter ablation is recommended. Success rates for ablation range from 85% to 95%, with higher success observed in patients with ILPVT. [10, 11, 12, 13, 14].

Previous studies have demonstrated changes in QRS morphology in recurrent patients with idiopathic left posterior ventricular tachycardia. Zhang et al. recently reported that upper septal ventricular tachycardia is identified in recurrent patients with the previous ablation of typical ILVT, which led to narrow QRS morphology. Other studies also had similar findings. Their mechanism of recurrence is reentry or a new focal origin. [15, 16, 17].

A pacemaker is a device that uses electrical impulses to increase or regulate the heart rate and /or rhythm when the patient's own intrinsic function of conduction or impulse generation is impaired. A temporary external pacemaker is located outside the body to regulate the heart rate and/ or rhythm for a temporary period. [1, 18, 19].

Patient also diagnosed with ARF. The incidence of initial cases of ARF is highest in children aged 5–14 years. The patient fulfills 1 major criterion, namely carditis, and 2 minor criteria, fever and leukocytosis accompanied by evidence of streptococcal infection in the form of ASTO (+) examination results. The patient was treated with Penicillin G Benzathine injection, acetosal and prednisone. Acetosal is given in the third weeks [1, 20, 21, 22, 23].

Both rheumatic valve disease and rhythm disturbances can cause heart failure in these patients. Congestive heart failure (CHF) is a clinical syndrome in which the heart is unable to pump enough blood to the body to

meet its needs, to dispose of systemic or pulmonary venous return adequately, or a combination of the two. The condition of heart failure will induce cardiogenic shock [23, 24].

One study published in the Journal of Pediatric Cardiology found that children with ARF had a higher incidence of ventricular tachycardia compared to children without ARF. Another study published in the Journal of Pediatrics found that children with ARF had a higher risk of developing ventricular tachycardia, especially if they had a history of recurrent streptococcal throat infections [25, 26].

Ventricular tachycardia is a rare complication of ARF, occurring in a small percentage of cases. While most children with ARF do not develop this condition, close monitoring for cardiac complications is essential. Studies report a 2.5% incidence of ventricular tachycardia in children with rheumatic fever, compared to 0.5% in those without, and a 3.4-fold increased risk for affected children. [21, 27].

In this patient, the ILVT was concomitant with ARF, as evidenced by the improvement in cardiac function following catheter ablation. Clinically, the patient showed significant recovery, and echocardiographic findings demonstrated an increase in ejection fraction from 39% to 63%, despite the valvular pathology remaining uncorrected.

CONCLUSION

Idiopathic LVT should be considered in the differential diagnosis of any SVT where it does not respond to common therapy of ventricular tachycardia and ECG shows characteristic features. Idiopathic LVT can occur concomitant to another heart abnormality such as ARF. Knowing about this rare case but easily treatable, physicians could prevent morbidity and mortality in them.

Declaration by Authors

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