

Congenital Junctional Ectopic Tachycardia in a 6-month-old boy treated with Ivabradine: A Case Report

Dinesh Upreti¹, Samir Shakya¹, Ratna Mani Gajurel¹, Hemant Shrestha¹, Sanjeev Thapa¹, Manju Sharma¹

¹ Department of Cardiology, Manmohan Cardiothoracic Vascular and Transplant Centre, Institute of Medicine, Maharajgunj, Nepal

Corresponding Author: Samir Shakya

Department of Cardiology, Manmohan Cardiothoracic Vascular and Transplant Centre, Institute of Medicine, Maharajgunj, Nepal.

Email: sammy_shakya@hotmail.com

ORCID ID NO: 0000-0001-8350-0665

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Abstract

Congenital junctional ectopic tachycardia (JET) is a rare tachyarrhythmia found in young infants and is often difficult to treat. JET is characterized by incessant tachycardia which causes ventricular dysfunction, heart failure and high mortality. Ivabradine is one of the newer antiarrhythmic drugs with special indications in decreasing heart rate in adults with chronic heart failure. It has shown promising role in congenital JET. However, there are no guidelines recommending ivabradine as a first-line agent for congenital JET. We report an infant with congenital JET who was successfully treated with ivabradine.

Keywords: Congenital, Junctional ectopic tachycardia, Ivabradine

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Introduction

Junctional tachycardia or Junctional ectopic tachycardia (JET) is tachyarrhythmias that arises from atrioventricular (AV) node and AV junction including bundle of His complex. Increased automaticity of the AV node/ His bundle is responsible for JET that is triggered by factors like mechanical trauma, stretch, ischemia, and circulating catecholamines.¹ It is a narrow complex tachycardia that can have retrograde atrial conduction in a 1:1 pattern or AV dissociation with variable conduction to the atria. JET can be broadly classified as congenital JET and postoperative JET.²

Congenital JET is a rare and usually incessant tachyarrhythmia that presents in the first few months of life. Untreated, congenital JET has been associated with ventricular dysfunction, heart failure and high mortality in infants. It has been associated with high mortality up to 34%.^{3,4} Amiodarone has most frequently been used as the first line drug in the management of JET.¹ Ivabradine, a novel drug used in chronic heart failure has been shown to be effective in congenital JET. Few studies have demonstrated the rewarding result of ivabradine in pediatric population.⁵⁻⁷ We report an infant with congenital JET presenting as incessant tachyarrhythmia who was successfully managed with ivabradine.

Case Report

A 6-month-old male child weighing 5.5 kg sought medical attention with complaints of coryza, cough, noisy and fast breathing associated with irritability for 2 weeks duration. He was found to have tachycardia (without fever) at another hospital and referred to our centre for cardiac evaluation. Electrocardiogram revealed narrow QRS complex tachycardia with a heart rate of 220 beats/min with evidence of AV dissociation (Figure 1A). Echocardiogram revealed a structurally normal heart with normal left ventricular (LV) ejection fraction of 65%.

In view of narrow complex tachycardia at rate of about 220 beats/min, it was decided to give adenosine. A brachial venous access was obtained, and intravenous adenosine was administered at a dose of 0.1 mg/kg, to which there was no response. This was followed up by 0.2 mg/kg and 0.3 mg/kg doses. A 12-lead electrocardiogram was recorded during adenosine administration. There was no change in the ventricular rate or rhythm with the additional doses as well. Congenital JET was strongly suspected in view of age of the child, narrow QRS complex tachycardia and AV dissociation. Amiodarone was started at bolus dose of 5 mg/kg over 20 mins followed by continuous intravenous infusion at 5 mcg/kg/min. The infusion dose was gradually increased to 20 mcg/kg/min over the next 24 hours.

The ventricular rate decreased upto 160 beats/min, during which AV dissociation was more prominent (Figure 1B). QT interval was being regularly monitored during amiodarone infusion and was within upper limit of normal.

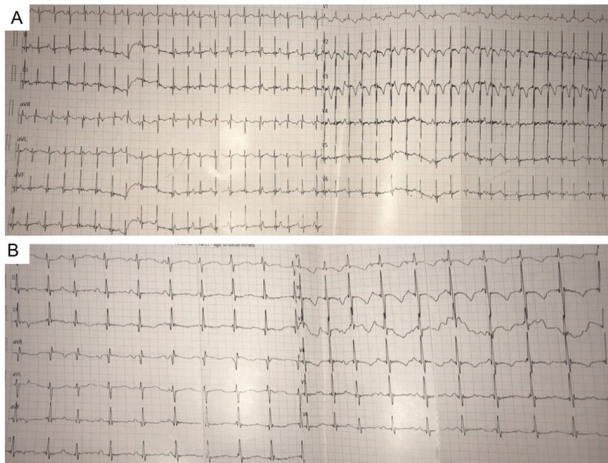


Figure 1. (A) Twelve lead ECG (sweep speed 25 mm/sec) showing narrow complex tachycardia at rate of 220 beats/min, with AV dissociation and ventricular rate faster than atrial rate suggestive of junctional ectopic tachycardia. (B) ECG (sweep speed 50 mm/sec) showing slower heart rate (160 beats/min) and prominent AV dissociation after amiodarone infusion. AV, atrioventricular; ECG, electrocardiogram.

In view of persistent JET, ivabradine was started at a dose of 0.1 mg/kg per dose twice daily. After 2 hours of the first dose of ivabradine, sinus rhythm was established with a heart rate of 120–140 beats/min (Figure 2A). Amiodarone was gradually tapered off over the next 24 hours. The baby remained in sinus rhythm at the same dosage of ivabradine alone for the next 3 days, following which he was discharged on ivabradine monotherapy (same dose). There was no occurrence of side effects like hypotension or bradycardia.



Figure 2. Twelve lead electrocardiograms of the infant, (A) obtained 2 hours after ivabradine therapy (sweep speed 50 mm/sec) and (B) on follow up after 1 week (sweep speed 25 mm/sec). Both electrocardiograms show sinus rhythm with well controlled ventricular rate.

At two weeks follow-up period, the baby remained in sinus rhythm with heart rate of 90-100 beats/min (Figure 2B) and the ventricular function was normal.

Discussion

The mechanism of tachyarrhythmia in congenital JET is believed to be due to an enhanced automaticity of the AV nodal region which in turn is due to slow, inward sodium current (the funny sodium current). This funny sodium current enters the cells through specific hyperpolarization-activated cyclic nucleotide-gated (HCN4) sodium channels. Overactivity of the HCN4 channels is proposed to be the molecular mechanism of this tachyarrhythmia, and ivabradine, as a selective inhibitor of the HCN4 channels, could potentially block this molecular mechanism.¹

The pharmacological treatment of congenital JET is difficult and often requires combination of anti-arrhythmic medications like amiodarone, digoxin, flecainide and propranolol.^{2,4} Few case reports have been published where ivabradine alone or in combination with other anti-arrhythmics has shown rewarding response in the treatment of this tachyarrhythmia. Kothari et al reported two siblings with JET and ventricular dysfunction who were not controlled despite multiple antiarrhythmic agents, and both of them responded dramatically to ivabradine.⁶ All other drugs could be gradually withdrawn, and both these patients were managed with ivabradine as monotherapy. In our case, there was a rapid conversion of JET to sinus rhythm after adding ivabradine to maximum dose of amiodarone infusion. Patient could be discharged at a controlled sinus rate on ivabradine monotherapy with maintenance of sinus rhythm on short term follow up.

Ivabradine was used as an adjunctive therapy by Dieks et al in five children with poor control on amiodarone and beta-blocker. It resulted in conversion to sinus rhythm in four and good rate control in the other. There were no significant adverse effects attributable to ivabradine therapy during a follow-up period of seven months.⁸ Ivabradine was successfully used to control congenital JET in a preterm neonate with congenital JET by Asfour et al after the rhythm could not be reverted with combination of propranolol and amiodarone.⁵ Devprasad et al reported successful use of ivabradine as monotherapy in a preterm neonate with congenital JET.⁷ Despite the poor response to other aforementioned medications, ivabradine has not been utilized as the first-line therapy in neonates and young infants with congenital JET.

Conclusion

Our report suggests that ivabradine can be effectively and safely used in the management of infants with congenital JET. It can be considered as an effective and safe alternative to the first line drug, amiodarone. However, further studies are warranted, especially to determine the continued efficacy and safety of ivabradine over longer periods.

Declaration of patient consent

The authors certify that they have obtained consent from the parent for publication of clinical information and investigation report.

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Conflicts of interest

There are no conflicts of interest.

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