Treatment of Permanent Junctional Reciprocating Tachycardia in a Preterm Neonate: Case Report

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Abstract

Keywords

- fetal tachycardia
- permanent junctional reciprocating tachycardia
- neonate
- preterm
- antiarrhythmic agents

Permanent junctional reciprocating tachycardia (PJRT) is most often observed in infants and children and can lead to incessant tachycardia. PJRT is usually refractory to medical treatment. Tachycardia may infrequently occur in the fetus in which case fetal tachycardia transplacental treatment should be started immediately. Term delivery is recommended for fetuses with tachycardia in the absence of significant clinical compromise to avoid complications of preterm birth. Herein, a 36-week preterm neonate presented with PJRT. He had tachycardia in the fetal period and was treated with digoxin, amiodaron, and esmolol therapy after birth without undergoing the catheter ablation procedure.

Introduction

Permanent junctional reciprocating tachycardia (PJRT), first defined by Coumel et al¹ in 1967, is an infrequent form of re-entrant supraventricular tachycardia (SVT). PJRT is most often observed in infants and children and can cause incessant tachycardia at a rate between 120 and 250 beats/min. It was noted that electrocardiograms (ECG) show inverted P waves in inferior leads II, III, and augmented vector foot (AVF).² If the persistent tachycardia is not resolved, it may lead to congestive heart failure and tachycardia-induced cardiomyopathy.^{3,4} Tachycardia may infrequently occur in the fetal period that may lead to heart failure and hydrops fetalis.⁵ PJRT is usually refractory to medications and electrical cardioversion; thus, catheter ablation procedures may be considered as an alternative option. Digoxin, amiodarone, flecainide, or β -blockers are the most commonly used drugs in the medical therapy of PJRT;

however, rare side effects from using these medications can be observed.^{2–4,6,7} Term delivery is recommended for fetuses with SVT in the absence of significant clinical compromise to avoid complications of preterm birth.⁸ If PJRT is detected in the fetal period, transplacental treatment with digoxin, sotalol, or flecainide should be initiated to prevent heart failure.⁵

In this article, a case was presented of a preterm neonate diagnosed with PJRT in the fetal period and treated with medical therapy.

Case Report

A 24-year-old pregnant female patient with a fetus at 36 weeks of gestation was referred to our pediatric cardiology unit due to sustained fetal tachycardia. The routine examination of mother and fetus until 36th gestational week during the pregnancy was normal. Fetal

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echocardiography revealed SVT with a fetal heart rate of 220 beats/min with 1:1 atrioventricular (AV) conduction. The ultrasonographic examination showed no manifestations of hemodynamic compromise, such as hydrops fetalis, cardiomegaly, or AV valve regurgitation. However, because the tachycardia was incessant (>50% of the time), the mother was recommended to begin sotalol therapy at 160 mg twice a day as transplacental treatment. Term delivery of the fetus was also recommended. Despite initiation of sotalol therapy, the fetal tachycardia continued incessantly. No maternal adverse events were observed during the transplacental therapy; however, recommendations to continue sotalol were not followed. Thus, a male baby weighing 2,920 g was delivered by cesarean section at the 36th gestational week, 24 hours after the initiation of sotalol therapy. He presented with hypotonia and severe respiratory distress on his first physical examination after birth. A heart rate of 210 beats/min, blood pressure of 51/30 mm Hg, respiratory rate of 72 breaths/min, grunting, and body temperature of 36.8°C were recorded. Bilateral rales were detected on lung auscultation. Hepatomegaly was also detected on abdominal examination. An analysis of arterial blood gases revealed pH of 7.09, pCO₂ of 72 mm Hg, pO₂ of 28 mm Hg, lactate of 4.6 mmol/L, HCO₃ of 16.4 mmol/L, and a base excess of -8.9 mmol/L. The patient was placed on mechanical ventilator support due to respiratory failure. A chest X-ray revealed pulmonary infiltration in the right lower lobe. Complete blood count, biochemical tests, and C-reactive protein were normal. An ECG showed narrow QRS tachycardia with a heart rate of 215 beats/min, prolonged RP interval, 1:1 AV conduction, and inverted P waves in leads II, III, and AVF (→ Fig. 1). A transthoracic echocardiographic examination revealed that there were no significant structural cardiac anomalies and

systolic functions was normal. The patient was diagnosed with PJRT based on these findings. Adenosine was administered intravenously in incremental doses of 100 µg/kg, 200 µg/kg, and 300 µg/kg. The tachycardia was sustained despite the short-term response following each dose of adenosine. The patient also responded to synchronized cardioversion for a short period of time, but the tachycardia resumed within seconds. A loading dose of amiodarone 5 mg/kg was initiated over 2 hours. During the second hour of amiodarone loading, the heart rate returned to sinus rhythm with 140 to 150 beats/min; however, hypotension and cardiac arrest occurred during this period but was resolved with short-term resuscitation. Amiodarone was discontinued and digoxin was initiated at a maintenance dose. A dopamine, dobutamine, and norepinephrine infusion was initiated upon the development of hypotension that led to a decrease in systolic functions on transthoracic echocardiography. After the sinus rhythm continued for 2 hours, PJRT started again at a rate of 220 beats/min. Based on this, an esmolol infusion was initiated at a rate of 30 µg/kg/min; however, it was discontinued due to the occurrence of cardiac arrest within the first hour of infusion. Amiodarone was restarted at a maintenance dose of 5 µg/kg/min without a loading dose owing to the persistence of tachycardia at 30 hours of life. At the second day, an esmolol infusion was added to the treatment again at a dose of 30 µg/kg/min due to persistence of tachycardia, and amiodarone was titrated to 15 mg/kg/day that was the peak infusion dose. The sinus rhythm was restored at 66 hours of life. The esmolol infusion was stopped at 92 hours of life. The tachycardia did not relapse, and the amiodarone infusion rate was reduced to 7 µg/kg/min. The patient's trachea was extubated on day six of life when the heart rate was controlled by a combination of

Fig. 1 Electrocardiogram showing a narrow QRS complex tachycardia with inverted P waves in II, III, and AVF (arrows). AVF, augmented vector foot.

Table 1 Prenatal, natal, and postnatal properties of the present patient

Prenatal properties	<ul style="list-style-type: none"> • A fetus at 36 weeks of gestation with incessant fetal tachycardia • Fetal echocardiography: SVT with a fetal heart rate of 220 beats/min and 1:1 AV conduction without hydrops or fetal compromise • Treatment: Sotalol at 160 mg twice a day
Natal properties	<ul style="list-style-type: none"> • A male baby weighing 2,920 g was delivered by cesarean section at the 36th gestational week, 24 h after the initiation of sotalol therapy
Postnatal properties	<ul style="list-style-type: none"> • ECG: narrow QRS tachycardia with a heart rate of 215 beats/min, prolonged RP interval, 1:1 AV conduction, and inverted P waves in leads II, III, and AVF • Echocardiography: No cardiac anomalies and normal systolic functions. • First day <ul style="list-style-type: none"> - PJRT, refractory to repeated adenosine and cardioversion - Digoxin started - Resolving after a loading dose of amiodarone 5 mg/kg, however, it was discontinued due to the occurrence of cardiac arrest - Esmolol was started at a rate of 30 µg/kg/min; however, it was discontinued due to the occurrence of cardiac arrest • Second day <ul style="list-style-type: none"> - Amiodarone was restarted at a maintenance dose of 5 µg/kg/min and increased to 15 mg/kg/day - Esmolol infusion was added to amiodarone therapy at a dose of 30 µg/kg/min due to persistence of tachycardia • Third day <ul style="list-style-type: none"> - The sinus rhythm was restored at 66 h of life • Fourth day <ul style="list-style-type: none"> - The esmolol infusion was stopped - Amiodarone infusion rate was reduced to 7 µg/kg/min • Sixth day <ul style="list-style-type: none"> - Extubated, the heart rate was controlled by an amiodarone and digoxin • Eighth day <ul style="list-style-type: none"> - The patient died due to Candida sepsis

Abbreviations: AV, atrioventricular; ECG, electrocardiogram; PJRT, permanent junctional reciprocating tachycardia; SVT, supraventricular tachycardia.

amiodarone and digoxin. Enteral feeding was initiated; however, the patient died on day eight of life due to septic shock caused by an invasive Candida infection (► **Table 1**).

Discussion

Fetal tachycardia is defined as heart rate more than 180 beats/min. According to its persistence, it can be classified as “sustained” if the tachycardia is present for more than 50% of the examination time or “intermittent” if periods of tachycardia alternate with predominantly normal heart rate. According to the electrophysiological appearance of the ECG and activity of the heart, the tachyarrhythmias can be classified as sinus tachycardia, atrial tachycardia (atrial flutter and atrial ectopic tachycardia), conduction system tachycardia (AV re-entry tachycardia [AVRT], junctional tachycardia, AV nodal re-entry tachycardia [AVNRT]), and ventricular tachycardia. From a practical, clinical standpoint, the classification of tachyarrhythmias can be summarized into (i) sinus tachycardia; (ii) SVT, which includes atrial and conduction system types; and (iii) ventricular tachycardia. AV re-entry tachycardia accounts for 90% of fetal and neonatal forms of SVT.^{9,10} PJRT is a subtype of AVRT and is a rare form of refractory and persistent SVT. PJRT generally occurs in children and infants and accounts for 1% of SVT in this age group.² During PJRT-induced tachycardia, the cardiac impulse is conducted in an antegrade manner through the AV node and His-Purkinje system, and is returned retrogradely via a slow, decremental concealed accessory pathway,

which is frequently located near the ostium of the coronary sinus. The arrhythmia is usually incessant from birth or infancy, although it may not be recognized until adulthood.^{2,4,11} PJRT presents incessant tachycardia at a rate between 120 and 250 beats/min. The ECG inscribes inverted P waves in leads II, III, and AVF; longer RP interval than P interval consistent with slow retrograde conduction; slightly slower heart rates than typical SVT, narrow or normal QRS. It is common for the AV conduction to be sensitive to adenosine treatment with tachycardia terminating with an AV or ventriculoarterial block but then recurring shortly afterwards.^{2,11} ECG is of great importance for differential diagnosis. When evaluating the ECG of narrow complex tachycardia, the diagnosis is narrowed to the typical form of AVNRT, AVRT, atrial tachycardia, sinus tachycardia, atrial flutter, and PJRT. In sinus tachycardia, heart rates may be mildly elevated (160–180 beats/min) or as high as 200 beats/min, A:V ratio is 1:1, and RP/PR ratio is high. If A:V ratio is more than 1:1, atrial tachycardia, atrial flutter, and multifocal atrial tachycardia are considered. If A:V ratio is 1:1, typical AVNRT, orthodromic AVRT, and PJRT are considered. In typical AVNRT and orthodromic AVRT, RP interval is short; whereas in PJRT, RP interval is longer than PR interval. Heart rate ranges between 220 and 280 beats/min in AVRT, 440 and 450 beats/min (atrial rate) in atrial flutter, and 180 and 220 beats/min in PJRT. There are three or more distinct P wave forms in multifocal atrial tachycardia; however, inverted P waves in the inferior lead in PJRT.^{2,9,12} The present case had findings of incessant narrow

QRS tachycardia with a heart rate of 215 beats/min, 1:1 AV conduction, prolonged RP interval and inverted P waves in the inferior leads on ECG, and an intravenous adenosine bolus terminated the tachycardia, which then spontaneously restarted after 5 seconds of sinus rhythm. Therefore, the patient was diagnosed with PJRT.

PJRT presents with symptoms in 69% of patients and the remaining 31% of patients are diagnosed with it, incidentally, through physical examinations or an ECG. If the persistent tachycardia is not treated, it may lead to cardiac dysfunction and tachycardia-induced cardiomyopathy. Symptoms of congestive heart failure are more common in infants compared with children older than 1 year, where incidence of heart failure is 19%. Fetal tachycardia is presented in 7% of patients. The rate of tachycardia is higher in infants compared with older children. Tachycardia-induced cardiomyopathy is observed in ~18% of patients. Accessory pathways of PJRT are generally isolated and not associated with structural heart defects.^{3,4} The patient in this case had symptoms of heart failure, but this may be due to persistent tachycardia from the fetal period. However, no structural heart defects or cardiomyopathy were detected.

PJRT can be difficult to treat and it is often refractory to drug therapy. In younger children, antiarrhythmic agents are generally the treatment of choice. Most of these infants and children require antiarrhythmic therapy with more than a single agent. However, if the tachycardia does not resolve with medical treatment, catheter ablation procedures may be required as an alternative option.^{2,4,6,13,14} In this specific case, although tachycardia was resolved completely on the sixth day of life without further need of the catheter ablation procedure in the patient, he then died due to septic shock. Intravenous adenosine is recommended as the first choice of treatment to terminate hemodynamically stable acute episodes of re-entrant SVT involving AV node. PJRT is usually refractory to adenosine.^{2,12} Although earlier studies have stated that using a combination of amiodarone, digoxin, and verapamil is the most effective regimen, currently, a combination of flecainide and amiodarone has been reported to be slightly more successful in the treatment of PJRT and reversing cardiomyopathy.² In a study by Kang et al³ antiarrhythmic medications were used for initial management in 76% of patients, while catheter ablation was used initially in only 10% of patients. They also reported that medications achieved clinical benefit in 64% of patients and complete resolution in 23% of patients, and 72% of all patients underwent catheter ablation procedures with a success rate of 90%. Using medication is more common in infants with 99% of them receiving medications compared with 60% of older patients. They concluded that resolution of PJRT in the absence of ablation therapy is difficult, and therefore, they recommend that the initial treatment for children and adolescents should be catheter ablation and medications should be used as the primary therapy for infants and small children.³ The tachycardia in this case was refractory to repeated adenosine administration and medical treatment was the first choice for the primary therapy of the patient, as he was a newborn.

Most of the fetal tachycardia due to ectopic focus resolves spontaneously and may no longer be observed when a scan

using echocardiography is performed. However, in a minority of cases, rhythm irregularities may represent a significant dysrhythmia, including conduction abnormalities in ~2.5% of the patients. If the fetal tachycardia is intermittent (<50% of the time) and with no hemodynamic compromise (e.g., no hydrops fetalis, no cardiomegaly, or no AV valve regurgitation), treatment may not be required. If the tachycardia is incessant (>50% of the time) or hemodynamic compromise is present, prenatal treatment should be commenced.⁹ It is recommended that if PJRT is diagnosed in the fetal period, transplacental treatment should be commenced as soon as possible to prevent congestive heart failure. Transplacental treatment should be initiated with a combination of sotalol and digoxin in nonhydropic patients, or flecainide, especially in case of fetal hydrops.⁵ Miyoshi et al¹⁵ reported that in fetuses with short ventriculoatrial SVT and no hydrops, the resolution rate was 46.7% with digoxin alone, 71.4% with digoxin plus sotalol, and 100.0% with digoxin plus flecainide. They concluded that protocol-defined transplacental treatment was effective and tolerable in 90% of fetuses with SVT and atrial flutter, and that severe adverse events which can occur in the fetuses including tachyarrhythmias may reoccur within the first 2 weeks after birth. During transplacental therapy, maternal adverse events are frequent and usually mild, but serious maternal adverse events such as AV block may rarely be observed. Therefore, the mother should be assessed by using echocardiography and ECG, and maternal serum levels of the antiarrhythmic agents should be measured during the transplacental therapy.¹⁵ The patient in this study did not show hemodynamic compromise during the fetal period; however, the fetal tachycardia was incessant. Therefore, we commenced sotalol to the mother as transplacental treatment. We did not observe any maternal adverse events during the short duration of maternal transplacental therapy.

A previous report highlighted a threefold increase in mortality rate in late preterm infants compared with term infants.¹⁶ Hinkle et al⁸ strongly proposed term delivery for fetuses with SVT in the absence of significant clinical compromise. Second-line or combination therapy should be considered for fetal SVT control so as to deliver at term to avoid complications of preterm birth; however, patients with refractory SVT are at an increased risk of premature delivery.⁸ Although transplacental treatment with sotalol and term delivery was recommended to the mother, the baby was born at the 36th gestational week. This proved to be a disadvantage in the case of complications related to preterm birth.

It was concluded that intravenous amiodarone may cause sinus bradycardia, AV block, hypotension, and cardiovascular collapse that mostly occur within the first 90 minutes of intravenous amiodarone infusion.¹⁴ It was also noted that esmolol (0.5 mg/kg rapid intravenous bolus followed by 50–200 µg/kg/min) can be used for the termination of neonatal SVT.¹² Rare side effects including bradycardia, decompensated heart failure, asystolic cardiac arrest, and heart block may be caused by esmolol administration.⁷ In this case, cardiovascular collapse occurred and hypotension developed at the second hour of administering the amiodaron loading dose, and cardiac arrest occurred within the first hour of low-

dose esmolol infusion. Amiodarone and esmolol treatment had to be temporarily discontinued due to these side effects. A few days later, these treatments were started again at lower doses and no side effects were observed.

In conclusion, term delivery should be recommended for fetuses with SVT in the absence of significant clinical compromise. In the case of fetal tachycardia, transplacental treatment should be commenced as soon as possible to prevent development of heart failure and hydrops fetalis. It is recommended that medical treatment should be administered for the primary therapy of PJRT in the neonatal period. Catheter ablation procedure may also be considered for infants who are refractory to medical treatment.

Conflict of Interest

None declared.

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