

Prenatal Management and Outcome of Junctional Ectopic Tachycardia and Hydrops

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Introduction

Fetal dysrhythmias are reported in approximately 1–2% of all pregnancies and are a relatively common reason for referral to fetal medicine centers¹⁻³. Fetal tachycardia is a serious condition in which the fetus is at risk of congestive heart failure and the subsequent development of hydrops¹⁻⁴. This situation is associated with significant morbidity and mortality¹⁻⁴. In general, the diagnosis and assessment of these dysrhythmias can be accurately made using high-resolution 2-D ultrasound using M mode and Doppler assessment of the relationship between the atrial and ventricular contractions⁵. Junctional ectopic tachycardia (JET) is a rare form of fetal tachyarrhythmia, which usually occurs in the setting of surgery for congenital heart disease⁶. In addition, a congenital variety of JET not related to surgery has also been described. While post-surgical JET has a mortality up to 14%, congenital JET has a mortality up to 34%⁶. The congenital form of JET has rarely been reported during the prenatal period^{7,8}. We describe a case in which the diagnosis was suspected in utero in a patient referred to our Department because of the presence of important fetal ascitis. The diagnosis was based on the evidence of moderate tachyarrhythmia episodes without 1:1 AV relationship and intermittent absence of atrial contraction wave consistent with atrioventricular (AV) dissociation, together with ductus venosus reverse diastolic wave indicating mild cardiac failure. JET diagnosis was confirmed after birth.

Case Report

A 32-year-old woman, gravid 1, was referred to our Department at 23 weeks' gestational age because of the presence of important fetal ascitis. On ultrasound examination (Voluson e8, GE), no heart or other structural anomalies were noted, fetal growth was adequate for

Keywords

Prenatal care / utilization; tachycardia; ectopic junctional; heart rate; hydrops fetalis.

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gestational age, and polyhydramnios as well as ascitis were observed, indicating fetal hydrops. Ventricular tachycardia episodes without 1:1 AV relationship and atrioventricular (AV) dissociation were observed (figure 1). In addition, a ductus venosus reverse diastolic wave was also noted. Fetal diagnosis was AV dissociation with either JET or ventricular tachycardia. Fetal JET was suspected on the basis of ventricular tachycardia episodes rarely exceeding 190 bpm without 1:1 AV relationship and atrioventricular (AV) dissociation, together with ductus venosus reverse diastolic wave and hydrops indicating cardiac failure. Thus, the fetus was diagnosed as having tachyarrhythmia, consistent with JET, and hydrops. The pregnant woman was admitted for an attempt of cardioversion with digoxin. After normal maternal ECG and electrolytes, treatment was started with oral digoxin 0.25 mg at 6 hourly intervals for a total of 2 mg, followed by a daily dose 0.25 mg at 8 hourly intervals. Once digoxin was within the therapeutic range (0.8-2 ng/ml), the patient was discharged and fetomaternal therapy monitoring was performed in an outpatient basis. Treatment control was based in maternal digoxinemia and ECG, as well as fetal ultrasonography twice a week. Blood digoxin level kept at therapeutic range during treatment, amniotic fluid volume returned to normal range, and fetal ascitis get reduced and remained stable, despite ductus venosus reverse diastolic wave indicating mild cardiac failure. Despite that response, at 28 weeks gestation we decided to give betametasone injections to promote lung maturation as prolongation of pregnancy was not guaranteed due to the general unfavorable prognosis and difficult management of JET or VT. At 34 weeks' gestational age, a cesarean section was performed due to the appearance of sudden and permanent fetal tachycardia despite digoxin therapy. A male weighing 2500 g with Apgar score 10/10/10 was born and the newborn was admitted for study at the Neonatology Department. At admission, neonatal ECG showed a regular narrow QRS tachycardia with AV dissociation, and the diagnose of JET was established (Figure 2). Treatment with adenosine was started to completely rule out an atrial flutter. As far as JET diagnosis was confirmed, we used propranolol only, and amiodarone associated to propranolol as well, but with poor outcome. Currently, the newborn is 9 month of age, presents a similar characteristic EKG and he is being treated with propranolol and flecainide. He shows a very good ventricular function (EF 72%) by echocardiography and an EKG holter recording with an average frequency 138 bpm.

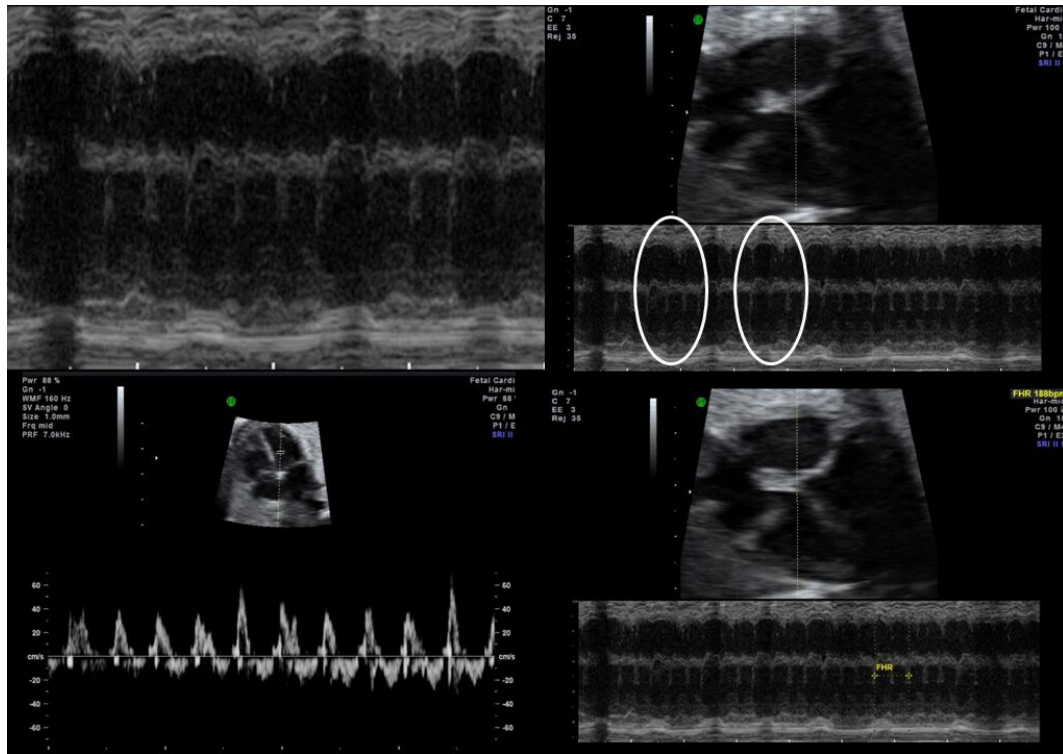


Figure 1 - A & B: AV dissociation with moderate ventricular tachycardia in M mode. **C:** intermittent absence of atrial contraction wave. **D:** ventricular tachycardia at 188 b/min in M mode.

Discussion

Cardiac arrhythmias are detected in approximately 1% of all fetuses^{1,2}. Most of these rhythm disturbances are the result of extrasystoles and are of little clinical significance^{1,2}. The incidence of fetal tachycardia is 1:10,000 to 1:25,000. Fetal tachyarrhythmias can result in the development of congestive heart failure and therefore prenatal intervention may be necessary^{1,2}. Junctional ectopic tachycardia is usually seen in children during the immediate postoperative period of congenital heart surgery. The congenital form is less frequent and has rarely been reported prenatally^{7,8}. Congenital JET is believed to be caused by an automatic ectopic focus in the AV junction³. In general, prenatal differential diagnosis between JET and VT on fetal echocardiogram is a challenge. In our case, the diagnosis of JET in utero was suspected on the basis of ventricular tachycardia episodes rarely exceeding 190 beats/min without 1:1 AV relationship and atrioventricular (AV) dissociation, together with ductus venosus reverse diastolic wave and ascitis indicating cardiac failure.

The optimal treatment of fetal tachyarrhythmias remains undetermined. The safety of the mother is of great concern when managing fetal tachycardia, as administration of antiarrhythmic drugs for intrauterine treatment may cause pro-arrhythmia and threaten the mother. Digoxin has been widely accepted as the mainstay of therapy because of its

safety and convenient administration^{1,2,5}. Flecainide, sotalol and amiodarone has also been used in the treatment of fetal tachyarrhythmias^{5,9,10}. However, more serious side effects on both mother and fetus have been a concern⁵.

The outcome in congenital JET shows that is a malignant tachyarrhythmia causing tachycardia-induced cardiomyopathy and sudden death⁶. Because of the limited efficiency of most antiarrhythmic medications in JET, the intrauterine management of such cases is difficult and should aim at maintaining adequate cardiac output to allow prolongation of the pregnancy until lung maturation is achieved^{7,8}. In addition, the presence of hydrops in our case suggested an unfavorable prognosis, despite the fact that, before digoxin therapy, tachyarrhythmia was episodic and ventricular rate rarely exceeded 190 beats/min.

In the case reported here, digoxin therapy was enough to control fetal heart rate, reduce fetal ascitis and keep amniotic fluid within the normal range, despite ductus venosus reverse diastolic wave indicating mild cardiac failure. Finally, at 34 weeks' gestation the fetus became non-responder to digoxin therapy and we decided to terminate pregnancy to avoid further fetal cardiac deterioration.

In conclusion, we report a case of complicated fetal JET that underwent transplacental fetal therapy with digoxin. Prenatal management of fetal arrhythmia is important to improve

Case Report

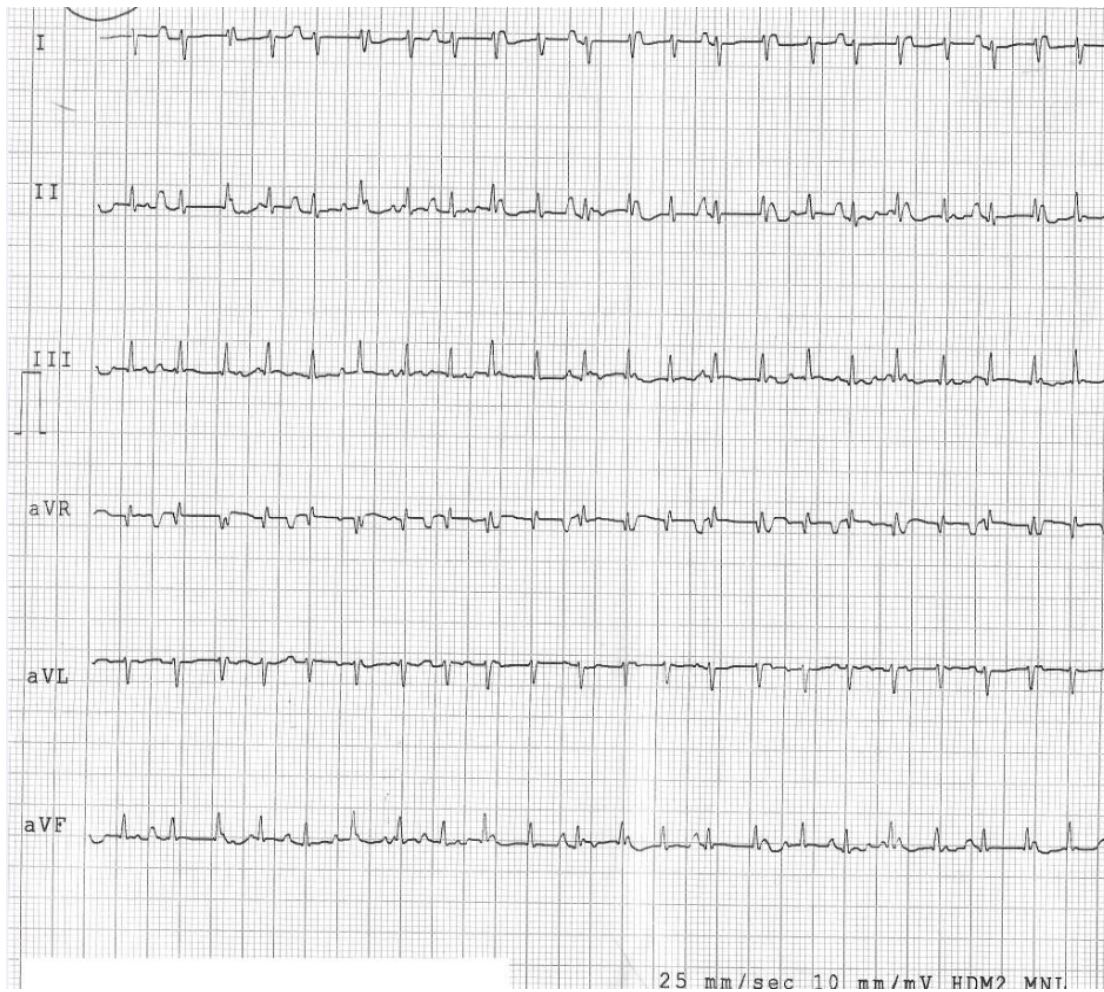


Figure 2 - Neonatal ECG showing a regular narrow QRS tachycardia with AV dissociation.

the outcome of affected fetus. Accurate prenatal diagnosis is crucial to the selection of the appropriate prenatal and postnatal treatments. Fetal JET is a rare condition and prenatal diagnosis remains a challenge, however differential diagnosis of fetal tachyarrhythmia should include this disorder.

Potential Conflict of Interest

No potential conflict of interest relevant to this article was reported.

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Study Association

This study is not associated with any post-graduation program.

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