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Fetal dilated cardiomyopathy caused by persistent junctional reciprocating tachycardia

J. CORNETTE*, A. D. J. TEN HARKEL† and E. A. P STEEGERS*

**Department of Obstetrics and Gynecology, Division of Obstetrics and Prenatal Medicine and †Department of Pediatric Cardiology, Erasmus MC, Rotterdam, The Netherlands*

KEYWORDS: amiodarone; dilated cardiomyopathy; ductus venosus; persistent junctional reciprocating tachycardia

ABSTRACT

Ultrasound examination of a fetus at 32 weeks' gestation revealed dilated cardiomyopathy and a heart rate of 170 beats per minute. Prenatally, this mild tachycardia was not primarily suspected to be the cause of the myocardial changes. Postnatal electrocardiography revealed a persistent junctional reciprocating tachycardia (PJRT) and the diagnosis of tachycardia-induced cardiomyopathy (TICM) became apparent. After conversion to a sinus rhythm under digoxin and amiodarone, the cardiac changes regressed. PJRT is a rare form of supraventricular tachycardia. The prenatal findings in the condition have previously been described retrospectively, but it can only be diagnosed postnatally by its characteristic electrocardiographic properties. This case indicates that TICM can occur at lower heart rates than previously assumed. Even severe prenatal cardiomyopathy may be reversible once sinus rhythm has been restored. Copyright © 2009 ISUOG. Published by John Wiley & Sons, Ltd.

CASE REPORT

A 29-year-old woman, gravida 2 para 1, was referred at 32 + 1 weeks' gestation for cardiomegaly and a fetal heart rate (FHR) of 170 beats per minute (bpm). The mild tachycardia had been noticed 3 weeks earlier but no action had been taken until the subsequent examination that led to referral to our unit. Echocardiography revealed severely dilated hypocontractile left and right ventricles with an increased cardiothoracic index (Figure 1). There was mild supraventricular tachycardia (SVT) of 165–170 bpm with 1 : 1 conduction and ventriculoatrial (VA) interval (240 ms) exceeding the atrioventricular (AV) interval (110 ms). Severe tricuspid regurgitation was observed and examination of the ductus venosus (DV) demonstrated a raised venous pulsatility index but positive a-wave. Further structural ultrasound examination only revealed mildly echogenic bowel, possibly indicating a viral origin. There was no sign of fetal hydrops.

These findings led to the diagnosis of severely dilated cardiomyopathy (DCM) with poor systolic function and early signs of fetal decompensation. The mild fetal tachycardia appeared to be secondary to the DCM rather than the causative condition. As such no antiarrhythmic therapy was considered. The woman was admitted for monitoring and steroids were administered for lung maturation.

The next day, the fetus showed progressive signs of decompensation with reversal of the a-wave in the DV. We opted to deliver the fetus despite the gestational age, in order to allow neonatal care before further deterioration. A male infant weighing 2060 g was delivered by Cesarean section at 32 + 2 weeks in good condition.

Echocardiography confirmed the DCM with a fractional shortening of 10% and a left ventricular end-diastolic diameter far above the 95th percentile (Figure 2). Electrocardiography revealed a persistent junctional reciprocating tachycardia (PJRT) (Figure 3). Cardioversion to restore a sinus rhythm was unsuccessful, as was treatment using propanolol, flecainide and propafenone. After several weeks a sinus rhythm was obtained under a combination of digoxin and amiodarone administration. Heart function was further supported by diuretics and captopril. After pharmacological cardioversion, a progressive regression in ventricular dilatation occurred with improvement in systolic and diastolic function, and after 11 weeks the infant was discharged in good cardiac condition. Follow-up at 5 months showed a left ventricular diameter at the upper limits of normality and a shortening fraction of

Correspondence to: Dr J. Cornette, Department of Obstetrics and Gynecology, Division of Obstetrics and Prenatal Medicine, Erasmus MC-University Medical Centre Rotterdam, Room SK 4161, Dr. Molewaterplein 60, 3015 GJ Rotterdam, The Netherlands (e-mail: j.cornette@erasmusmc.nl)

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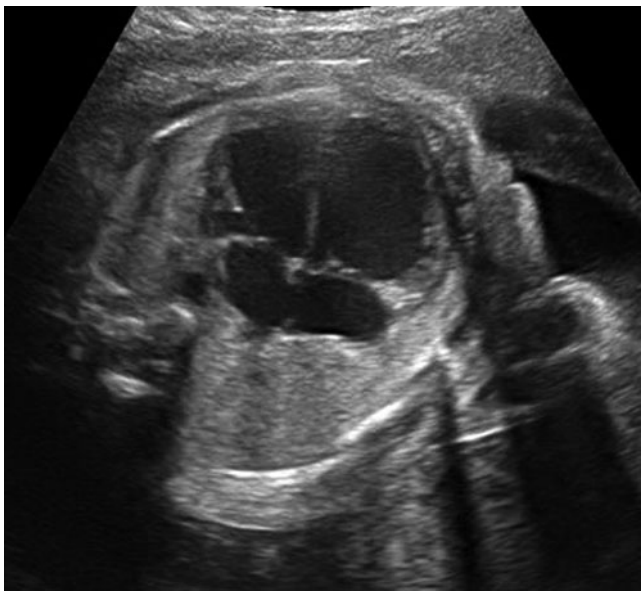

Figure 1 Prenatal ultrasound image at 32 weeks' gestation in a case of fetal dilated cardiomyopathy; note the severe dilatation of both ventricles and increased cardiothoracic index.

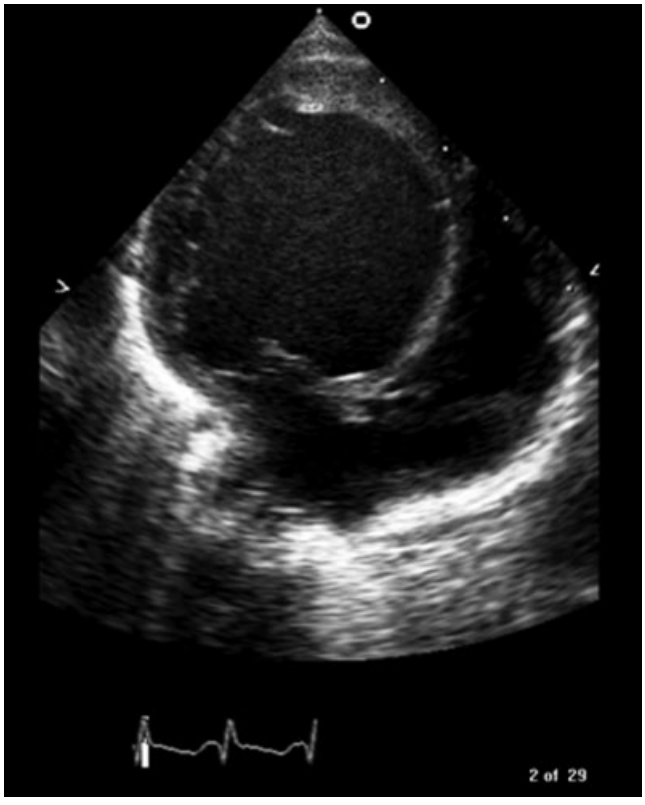

Figure 2 Ultrasound image shortly after birth, showing the very prominent dilatation of the left ventricle.

37% (Figure 4). Diuretics and digoxin could be discontinued at 9 months, while amiodarone and captopril were maintained on a low dose. At 17 months the left ventricular size and function remained stable and medication was stopped.

DISCUSSION

Fetal DCM is a rare diagnosis, occurring in 0.2% of high-risk pregnancies. It is characterized by dilatation and impaired systolic function of one or both ventricles, and in prenatally detected cases overall mortality is as high as 70%[1]. The underlying causes are heterogeneous, but some of them can be identified antenatally allowing treatment to be attempted.

Sustained tachyarrhythmias can produce significant myocardial remodeling, known as tachycardia-induced cardiomyopathy (TICM)[2,3]. The degree of cardiac remodeling appears to correlate with the rate and duration of the tachycardia, and rate control often leads to recovery.

The normal range for FHR is 110 to 160 bpm. Fetal tachycardia is defined as a sustained heart rate above 180 bpm[4]. Fetal TICM has been described with all types of SVT including PJRT, but is mostly associated with an FHR above 220 bpm[4–6].

In adults, DCM can also induce arrhythmias, and sinus tachycardia can occur as an attempt to compensate for the systolic dysfunction by a small increase in rate[7]. While not specifically described in prenatal medicine, one might expect a similar mechanism in the fetus. However, prenatal discrimination between causation and result is sometimes impossible.

PJRT is a rare form of re-entrant SVT, with an accessory retrograde pathway with slow conduction. This leads to a characteristic long RP interval exceeding the PR interval. There is a typical 1 : 1 conduction[6,8,9]. While most SVTs have intermittent episodes of normal sinus rhythm, PJRT is commonly incessant.

In the fetus, the AV relationship in SVT is usually investigated by Doppler examination of the mechanical consequences of electrical conduction. PJRT can be suspected when the VA interval exceeds the AV interval, but atrial ectopic tachycardia and sinus tachycardia have a similar appearance[10,11]. At a FHR below 200 bpm, differentiation from sinus tachycardia is extremely difficult[10,12], and as such diagnosis of prenatal PJRT has always been retrospective.

PJRT is often very resistant to pharmacological therapy[6,8,9]. Our patient was finally converted to sinus rhythm by treatment with digoxin and amiodarone. There have also been some previous positive reports on the use of the latter antiarrhythmic drug in the treatment of PJRT[9,13]. When pharmacological management fails, radiofrequency ablation of the accessory pathway is usually effective[9].

A major role has been attributed to myocardial hypoperfusion in a hypothesis on the pathophysiological mechanisms of TICM[14]. In the fetus, an abrupt change to a pulsatile flow in the precordial veins is observed above a critical heart rate of 210 bpm, reflecting raised venous pressure[5]. Ventricular filling is then disturbed by a reduced filling time and impaired ventricular relaxation, and myocardial hypoperfusion occurs owing to a shortened diastole and a reduced perfusion gradient[14].

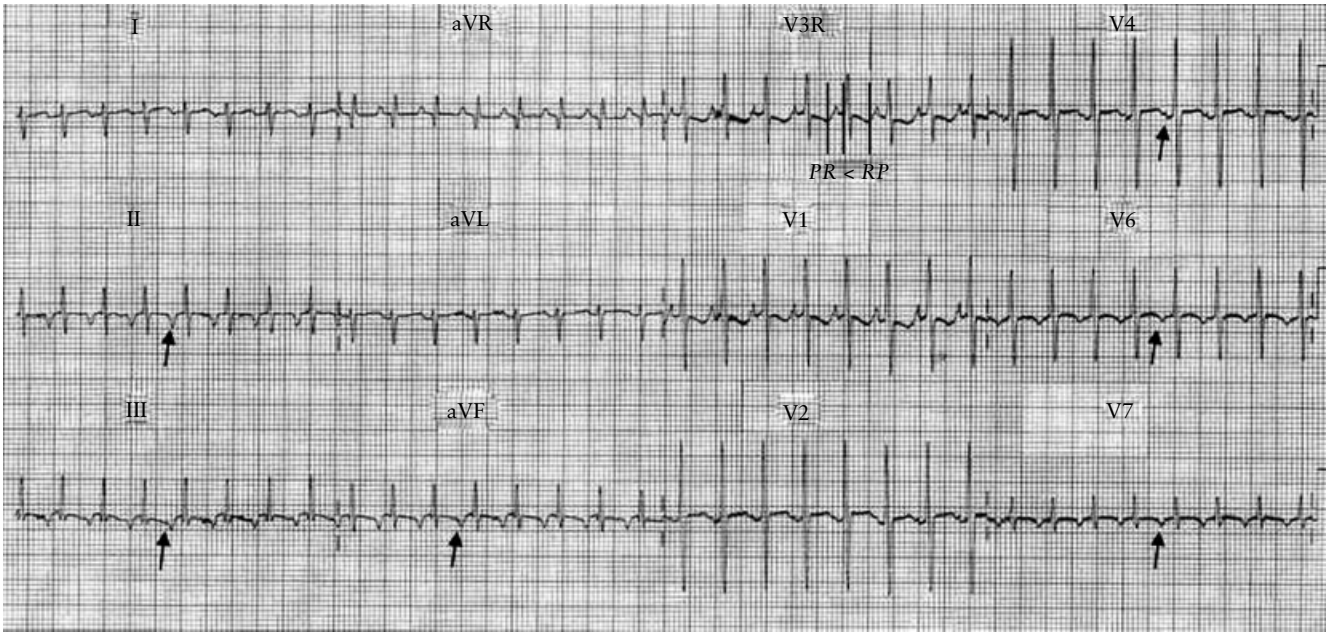

Figure 3 Neonatal 12-lead electrocardiogram with characteristic features of persistent junctional reciprocating tachycardia. There is a regular, narrow QRS tachycardia with negative P-waves in leads II, III, aVF and left lateral leads (arrows). There is a long RP interval (200 ms) which exceeds the PR interval.

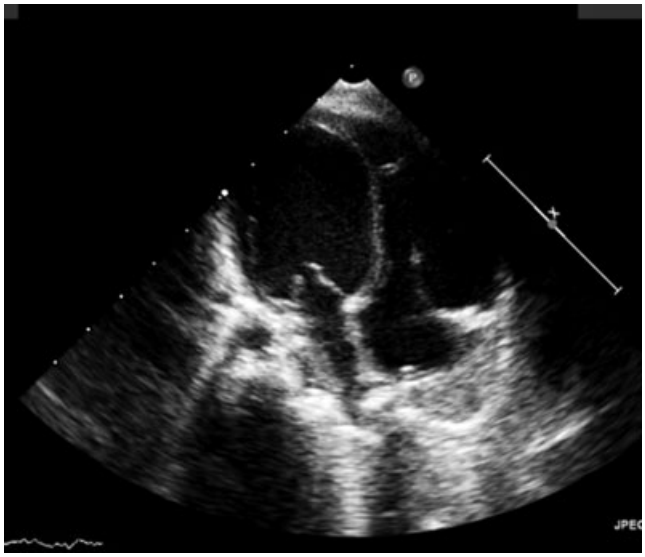

Figure 4 Ultrasound image of the infant at 5 months of age, showing the still-globular appearance of the left ventricle. The end-diastolic dimensions were at the upper limits of normality, and remained so at 17 months of age.

It is hypothesized that the increased oxygen demand from sustained tachycardia is met by a decreased coronary perfusion, resulting in myocardial injury with remodeling[14].

With ventricular dilatation, annular enlargement of the AV valve induces functional incompetence with valve regurgitation[14,15]. Altered flow patterns in the DV and inferior vena cava are believed to reflect the increased end-diastolic pressure caused by cardiomyopathy-induced changes in ventricular compliance and relaxation[14,15].

Although an FHR of 170 bpm is far below the critical level, it still induced a very prominent DCM in our case. We are not aware of any other cases of fetal TICM at this relatively low heart rate, but Chen *et al*. have reported a similar case of TICM due to a PJRT with a rate of 180–190 bpm[13]. The persistent character of the tachycardia in our case was probably the major contributor to the cardiac remodeling. This case also suggests that mechanisms other than myocardial hypoperfusion, such as depletion of energy stores, oxidative stress and intrinsic loss of myocardial reserve, may play a role in the pathophysiology of TICM[2,3].

Increased venous congestion can rapidly lead to decompensation, and timely intervention by either fetal or neonatal therapy is advisable. As the exact cause of the DCM was uncertain in our case, we opted for a planned delivery at 32 weeks when the fetus was still in good condition. Prematurity is unlikely to be a major concern at this gestational age and diagnostic and therapeutic options are more appropriate. Retrospectively, if the DCM had been diagnosed as TICM then transplacental therapy could have been attempted. However, as the PJRT proved difficult to manage postnatally, antenatal pharmacological cardioversion would most probably have failed.

In conclusion, it seems that fetal TICM can be induced at lower heart rates than previously assumed when the arrhythmia is persistent. Sustained rates above 165 bpm should lead to a suspicion of arrhythmia and be investigated by a detailed fetal heart scan. While diagnosis of DCM and fetal tachycardia is usually straightforward, distinguishing between causation and result can still be challenging. However, even severe prenatal cardiomyopathy may be reversible once sinus rhythm has been restored.

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