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Title: Decreased baseline variability on fetal heart rate pattern in a fetus with heterotaxy syndrome

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A short running title: Decreased FHR baseline variability

Title: Decreased baseline variability on fetal heart rate pattern in a fetus with heterotaxy syndrome

ABSTRACT

In a fetus with suspected heterotaxy syndrome, a decreased/absent baseline variability of fetal heart rate (FHR) pattern developed at gestational week (GW) 36^{-5/7} and continued for 5 days until birth at GW 37^{-2/7}, while repeat biophysical profile scorings with ultrasound were consistently unremarkable. This neonate weighing 2404 g with Apgar scores of 7 (1-min) and 8 (5-min) and umbilical arterial cord blood pH of 7.28 with base deficit of 3.9 mmol/L, showed a heart rate (HR) of 120 bpm for 3 hours after birth, but subsequently developed sinus bradycardia (84 bpm) unresponsive to crying. Isoproterenol initiated 9 hours after birth was effective in the increase of HR to 120 bpm in this neonate. Brain MRI at 16 days of age was unremarkable. The decreased/absent baseline variability of FHR pattern was speculated to have been caused by sinus node dysfunction, and not by reduced fetal oxygenation in this case.

Key words: cardiotocogram, congenital cardiac malformation, fetal arrhythmia, fetal heart rate pattern, isomerism

INTRODUCTION

In fetuses without known malformations affecting cardiac rhythm, the decreased fetal heart rate (FHR) baseline variability usually suggests fetal hypoxemia [1]. However, it is unclear whether fetuses with heterotaxy (right isomerism) syndrome and cardiac structural abnormalities are likely to exhibit decreased FHR baseline variability in the presence of adequate fetal oxygenation. Our case suggested that sinus node dysfunction in a fetus can cause a decrease in baseline variability of fetal heart rate pattern in the absence of fetal hypoxemia. This case presentation was conducted with the approval of the parents and the institutional review board of Hokkaido University Hospital.

CASE

A 39-year-old nulliparous pregnant Japanese woman experienced transient fetal bradycardia of 100 bpm and was referred to our institute at gestational week (GW) 22. Her fetus had suspected heterotaxy (right isomerism) syndrome and multiple cardiac malformations, including unbalanced atrioventricular septal defect, double outlet right ventricle with pulmonary stenosis, and common atrium on echocardiography. She developed hypertension (153/99 mmHg) and proteinuria (protein/creatinine ratio [mg/mg], 0.77) at GW 32^{-5/7} and GW35^{-4/7}, respectively. Although fetal well-being based on daily non-stress testing was assured until GW 36^{-4/7}, a decreased/absent baseline variability of FHR pattern developed at GW 36^{-5/7} (Fig. 1) and lasted until caesarean delivery at GW 37^{-2/7} for uncontrolled maternal hypertension (184/106 mmHg) while on 1500 mg methyldopa, whereas results of frequent biophysical scorings with ultrasound suggested consistent well-being in this fetus during the last 5 days *in utero*. This neonate, weighing 2404 g with Apgar scores of 7 (1-min) and 8 (5-min) and umbilical arterial cord blood pH of 7.28, PO₂ of 14.8 mmHg, PCO₂ of 50.7 mmHg, and base deficit of 3.9 mmol/L, showed a heart rate (HR) of 120 bpm for 3 hours after birth

followed by sinus bradycardia of 84 bpm unresponsive to crying (Fig. 2). Following continuous intravenous infusion of isoproterenol initiated 9 hours after birth, the HR increased to around 120 bpm and this rhythm continued even after discontinuation of isoproterenol at 6 days of age. This baby diagnosed as having unbalanced atrioventricular septal defect, double outlet right ventricle with pulmonary stenosis, common atrium and aorticocaval juxtaposition on postnatal echocardiography was confirmed to have right isomerism based on findings of bilateral right atrial appendages during modified Blalock-Taussig shunt surgery from median sternotomy performed at age of 28 days for the correction. This infant was doing well at age of 4 months when last seen.

DISCUSSION

It may be difficult to assess fetal well being based on electronic FHR pattern in some fetuses with cardiovascular malformations and/or cardiac dysfunction. The decreased/absent baseline variability of FHR pattern in the absence of drugs affecting the central nervous system usually suggests an unfavorable intrauterine environment with regard to fetal blood oxygenation [1]. Although fetuses with cardiac structural anomalies are likely to exhibit abnormal FHR pattern on electronic FHR tracing, with some cases requiring emergency cesarean section [2 – 4], decreased/absent baseline variability of FHR pattern is very rare in the absence of fetal hypoxemia; to our knowledge, there have been no reports documenting sustained decreased/absent baseline variability of FHR pattern in the presence of adequate fetal oxygenation even in fetuses with cardiac anomalies. This necessitated frequent biophysical scoring tests to ensure fetal well being. Chromosomal aberration may also contribute to non-reassuring fetal status based on FHR tracing [2]. However, as the present infant did not have any external malformation suggestive of chromosomal aberration and as the heterotaxy syndrome is generally considered not to be associated with chromosomal aberration, this infant did not undergo chromosomal analysis.

Anatomical abnormality of the sinus node and the sinus node dysfunction are frequently present in patients with isomerism of the atrial appendages [5 – 8]. Sinus node dysfunction is usually observed in patients with left isomerism. However, hypoplastic or absent sinus node is also seen sometimes in patients with right isomerism [6]. Based on the results of postnatal investigations and clinical course, the present case was speculated to have had at least transient functional abnormality of the sinus node, but not reduced fetal oxygenation. We speculated that immaturity of the cardiac conduction system as well as the sympathetic nerve system immediately after birth was responsible for this transient sinus node dysfunction. Alternatively, as twin sinus node and twin atrioventricular node present sometimes in patients with right isomerism [5, 6], change in dominancy of the sinus node was speculated to cause transient sinus node dysfunction in this patient.

This case suggested that fetal cardiac anomalies involving the sinus node can cause absent/decreased baseline variability of FHR pattern even in the presence of adequate fetal oxygenation.

Disclosure

The authors have no financial conflicts of interest to disclose concerning the manuscript.

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Figure legend

Fig. 1. Fetal heart rate patterns on electronic FHR tracing at GW 36^{-2/7} and 36^{-5/7}
Decreased variability with late deceleration was seen at GW36^{-5/7} in this case.

Fig. 2. ECG of a neonate on the day of birth

This ECG obtained 3.5 hours after birth shows sinus bradycardia with junctional escape beat of approximately 84 bpm. This heart rate did not increase in response to crying. However, the sinus node dysfunction was transient—the heart rate increased to around 120 bpm after starting continuous intravenous infusion of isoproterenol and normal rhythm was maintained even after discontinuation of isoproterenol at 6 days of age in this neonate.

Fig. 1

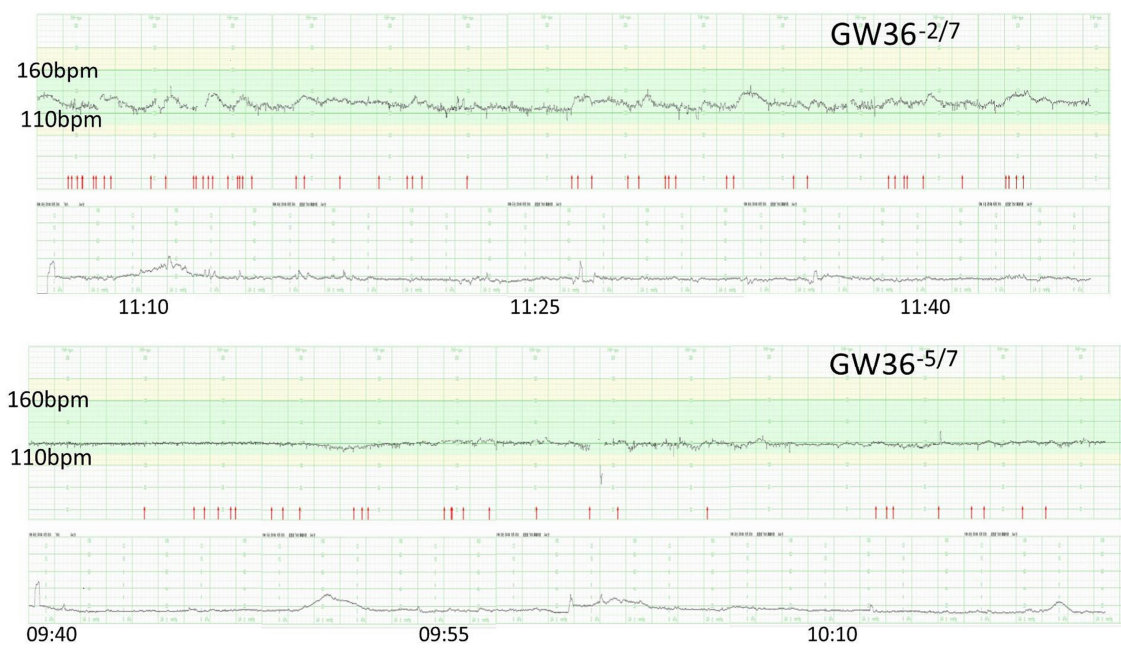


Fig. 2

