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# Prenatal Diagnosis of High Grade Atrioventricular Block with Polymorphic Ventricular Premature Contractions due to Congenital Long QT Syndrome Using Doppler Flow Recording

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**Abstract:** We herein report a case of fetal long QT syndrome with a series of cardiac arrhythmias resulting from a 2:1 atrioventricular block and high grade atrioventricular block with polymorphic ventricular premature contractions leading to ventricular tachycardia. These were detected in late gestation using the M-mode and simultaneous Doppler flow recording of the superior vena cava and the ascending aorta. Simultaneous Doppler flow recording of the superior vena cava and the ascending aorta yields more accurate measurements of the atrioventricular and ventriculo–ventricular intervals compared with the conventional M-mode. It is helpful for diagnosing arrhythmias, including various types of dissociation of atrioventricular conduction and ventricular arrhythmias.

**Key words**：**LQT, Bradycardia, Atrioventricular block, Ventricular tachycardia, Fetal echocardiogram**

## Introduction

Congenital long QT syndrome（LQTS）is a rare disorder that carries a high risk of life-threatening ventricular arrhythmias. In particular, cases with congenital LQTS diagnosed during the prenatal or early neonatal periods have a very poor prognosis.[1,2] A prenatal diagnosis of LQTS before birth is required so that intervention can be provided in the pre- and/or postnatal periods to avoid fetal and neonatal death.

With the introduction of fetal magnetocardiography（MCG），which detects faint magnetic signals generated from the fetal heart, there have been a few reports of the prenatal diagnosis of LQTS using MCG.[2,3] More recently, Fujimoto et al. reported the successful prenatal diagnosis of LQTS using fetal electrocardiography（ECG）.[4] However, fetal MCG and ECG are not widely available in the clinical setting. At present, there are not many institutions where such fetal MCG and ECG are available in Japan. Therefore, the detection of various arrhythmias due to LQTS using fetal echocardiography is required. There have been many reports regarding the prenatal diagnosis of severe arrhythmias, including bradycardia, atrioventricular（AV）block, ventricular tachycardia（VT）and their combinations due to LQTS that were detected using M-mode echocardiography.[5-8]

The simultaneous recording of blood flow velocity waveforms obtained from the superior vena cava（SVC）and the ascending aorta（AAo）using pulsed-Doppler ultrasonography（SVC/AAo Doppler flow recording）is one of the standard examinations for the diagnosis of fetal arrhythmias.[9,10] SVC/AAo Doppler flow recording is superior to the conventional M-mode, because of its better resolution to identify the beginnings of hemodynamic events corresponding to atrial and ventricular contractions,

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and it yields more accurate measurements of the AV and ventriculo–ventricular intervals. Using this method, fetal LQTS has been diagnosed by detecting 2:1 AV block.[11] However, to the best of our knowledge, there are no previous case reports of congenital LQTS with any other arrhythmias estimated using SVC/AA Doppler recording in a fetal echocardiogram. We herein report a case of fetal LQTS in which a series of cardiac arrhythmias resulting from a 2:1 AV block and high grade AV block with polymorphic ventricular premature contractions (VPCs) leading to VT were detected in late gestation using SVC/AAo Doppler flow and M-mode recordings.

## Case report

A 26-year-old Japanese pregnant patient, G1, P0-1-0-0, was referred to our department at 35+0 weeks gestation because of sustained fetal bradycardia of approximately 80 beats per minute. Her pregnancy had been uneventful. She had no history of syncope or

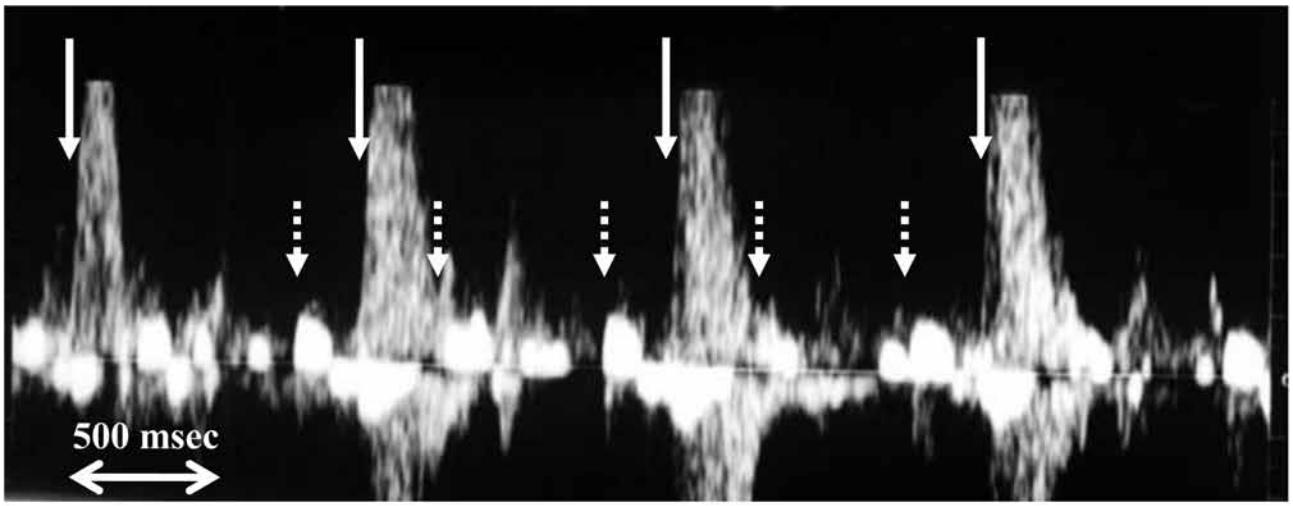


**Fig. 1.** Flow velocity waveforms of the superior vena cava (dotted arrows) and the ascending aorta (solid arrows) at 35+0 weeks' gestation indicating a 2:1 atrioventricular conduction block.

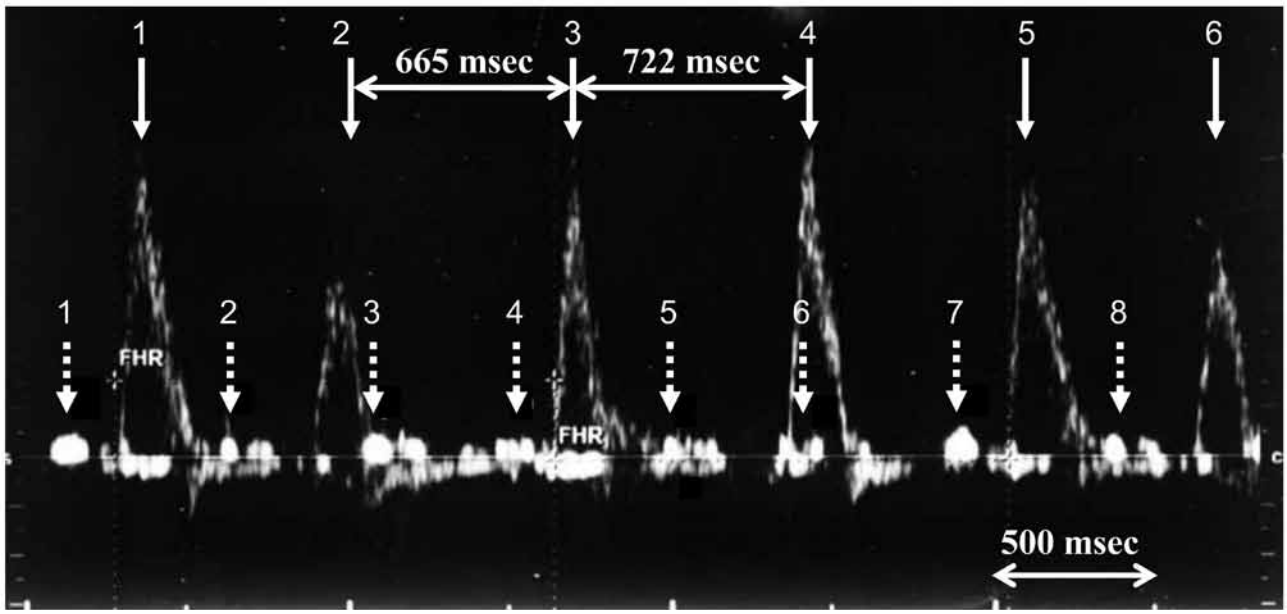


**Fig. 2.** Flow velocity waveforms of the superior vena cava (SVC) and the ascending aorta (AAo) at 36+0 weeks' gestation indicating a high grade atrioventricular conduction block and polymorphic ventricular premature contractions. The dotted arrows indicate the reverse flow waveforms of the SVC and solid arrows indicate the outflow waveforms of the AAo.

cardiac arrhythmias. On echocardiography, the fetal heart was slightly enlarged, with a cardio-thoracic area ratio（CTAR）of 43%. The SVC/AAo Doppler flow recording revealed a 2:1 AV block（Figure 1）. A maternal blood examination showed that the patient was negative for antinuclear antibodies, anti-SSA and anti-SSB antibodies. At 36+0 weeks' gestation, a SVC/AAo Doppler flow recording showed a dissociation of AV conduction except for the 1st and 7th *a* waves, indicating high grade AV block（Figure 2）. The shapes of the 2nd, 3rd and 4th *v* waves were variable, and the peak-to-peak intervals were different between the consecutive

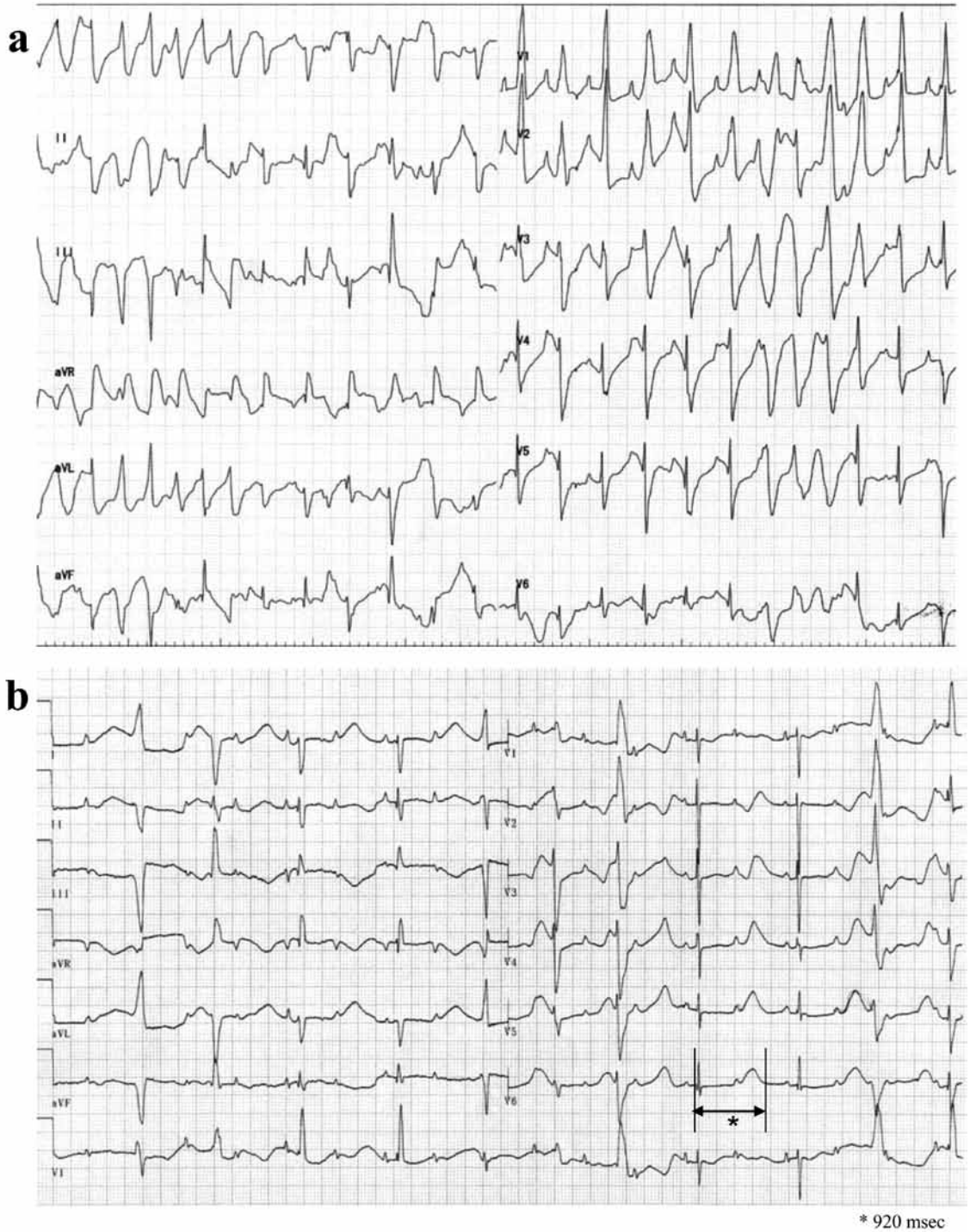


**Fig. 3.** Twelve-lead electrocardiograms at birth（a）showing polymorphic ventricular tachycardia, and after intravenous administration of xylocaine and amiodarone（b）showing high-grade atrioventricular block with polymorphic ventricular premature contractions with a marked prolongation of the QT interval to 920 msec（*）.

waveforms, indicating polymorphic VPCs. At 36+5 weeks' gestation, VT appeared, and fetal cardiomegaly progressed with a CTAR of 52%. An emergency cesarean section was performed at 36+5 weeks' gestation, and a 2,344-gram male infant was delivered. The Apgar scores were 4 at 1 minute and 4 at 5 minutes.

ECG at birth showed polymorphic VT, which disappeared following intravenous administration of xylocaine and amiodarone (Figure 3). A subsequent ECG revealed high grade AV block with polymorphic VPCs and a marked prolongation of the QT time to 920 msec. Based on these findings, LQTS was diagnosed, and temporary pacing was immediately performed. However, ventricular refractoriness was sustained, which indicated that the pacing was failing. Torsade de pointes appeared, and the patient died at one day of age. A genetic analysis revealed a *SCN5A* mutation (F1486del) that confirmed a diagnosis of LQTS type 3. [12] There were no mutations found in the genetic analysis of his parents.

## Discussion

In the present case, the SVC/AAo Doppler flow recording showed a high grade AV block with variable shapes in the AAo flow velocity waveforms and irregular peak-to-peak intervals indicating polymorphic VPCs. Polymorphic VPCs are considered to be the preparatory or preceding arrhythmia for polymorphic VT. In fact, in this case, the polymorphic VT recovered to polymorphic VPCs following the administration of xylocaine and amiodarone after birth. Polymorphic VT is expressed in each AAo flow with non-uniform morphology and irregular peak-to-peak intervals in Doppler recordings.[12] This suggests that polymorphic VPCs expressed by non-uniform and irregular peak-to-peak interval AAo flow are the result of the formation of arrhythmias which easily transition to polymorphic VT.

A 2:1 AV block or intermittent bradycardia with a 4:3 or 3:2 AV block in LQTS fetuses has been reported previously.[11, 14] However, there have been few case reports of congenital LQTS with high grade AV block. In the present case, the high grade AV block might have been caused by the simultaneously occurring functional conduction block and polymorphic VPCs because of the significantly long repolarization time. With regard to the electrophysiological mechanism of ventricular arrhythmias in LQTS, it has been suggested that single ectopic beats, couplets and the initial beat of polymorphic VT consistently arise as focal activity from a subendocardial site with shorter refractoriness, and are conducted to the surrounding deeper myocardial regions with longer refractoriness.[15] The prolongation of cardiac repolarization makes it difficult to create uniform electrical activities, resulting in the development of a functional conduction block.

SVC/AA Doppler recording is helpful to diagnose arrhythmia, using the flow waves in the echocardiogram to resemble electrical waves in ECG. Although such a method cannot measure the duration of the QT interval, it can detect high-risk LQTS with bradycardia, 2:1 or high-grade AV block and VT. Therefore, it may provide a useful alternative to MCG or ECG.

## Conclusion

We herein reported a case of severe congenital LQTS with 2:1 AV block, high grade AV block, polymorphic VPCs and VT. SVC/AAo Doppler flow recording is very helpful for prenatally diagnosing various arrhythmias, and for detecting high risk congenital LQTS.

## Conflict of Interest

There is no conflict of interest or competing financial interest in relation to this work.

## References

[1] Garson A, Jr., Dick M, 2nd, Fournier A, Gillette PC, Hamilton R, Kugler JD, van Hare GF 3rd Vetter V, Vick GW 3rd: The long QT syndrome in children. An international study of 287 patients. Circulation **87**: 1866-1872, 1993.

[2] Hosono T, Kawamata K, Chiba Y, Kandori A, Tsukada K: Prenatal diagnosis of long QT syndrome using magnetocardiography: a case report and review of the literature. Prenat Diagn **22**:198-200, 2002.

[3] Schmitz L, Burghoff M: Images in cardiovascular medicine. Magnetocardiography in a fetus with long-QT syndrome. Circulation **112**: e68-69, 2005.

[4] Fujimoto Y, Matsumoto T, Honda N, Tojo R, Furuya M, Kasai K, Saito S, Mochimaru F, Ishikawa Y: Prenatal diagnosis of long QT syndrome by non-invasive fetal electrocardiography. J Obstet Gynaecol Res **35**: 555-561, 2009.

[5] Hofbeck M, Ulmer H, Beinder E, Sieber E, Singer H: Prenatal findings in patients with prolonged QT

interval in the neonatal period. Heart **77**: 198-204, 1997.

[6] Ohkuchi A, Shiraishi H, Minakami H, Eguchi Y, Izumi A, Sato I: Fetus with long QT syndrome manifested by tachyarrhythmia: a case report. Prenat Diagn **19**: 990-992, 1999.

[7] Manning N, Anthony JP, Ostman-Smith I, Snyder CS, Burch M: Prenatal diagnosis and successful preterm delivery of a fetus with long QT syndrome. BJOG **107**: 1049-1051, 2000.

[8] Beinder E, Grancay T, Menendez T, Singer H, Hofbeck M: Fetal sinus bradycardia and the long QT syndrome. Am J Obstet Gynecol **185**: 743-747, 2001.

[9] Fouron JC, Proulx F, Miro J, Gosselin J: Doppler and M-mode ultrasonography to time fetal atrial and ventricular contractions. Obstet Gynecol **96**: 732-736., 2000.

[10] Fouron JC, Fournier A, Proulx F, Lamarche J, Bigras JL, Boutin C, Brassard M, Gamache S: Management of fetal tachyarrhythmia based on superior vena cava/aorta Doppler flow recordings. Heart **89**: 1211-1216, 2003.

[11] Furushima H, Chinushi M, Sato A, Aizawa Y, Kikuchi A, Takakuwa K, Tanaka K: Fetal atrioventricular block and postpartum augmentative QT prolongation in a patient with long-QT syndrome with KCNQ1 mutation. J Cardiovasc Electrophysiol. **21**: 1170-1173, 2010.

[12] Wang Q, Shen J, Splawski I, Atkinson D, Li Z, Robinson JL, Moss AJ, Towbin JA, Keating MT.: SCN5A mutations associated with an inherited cardiac arrhythmia, long QT syndrome. Cell **80**: 805-811, 1995.

[13] Takahashi K, Shiraishi H, Ohkuchi A, Matsubara S, Kuwata T, Yada Y, Izumi A, Suzuki M: Irregular peak-to-peak intervals between ascending aortic flows during fetal ventricular tachycardia in long QT syndrome. Ultrasound Obstet Gynecol **33**: 118-120, 2009.

[14] Lin MT, Hsieh FJ, Shyu MK, Lee CN, Wang JK, Wu MH: Postnatal outcome of fetal bradycardia without significant cardiac abnormalities. Am Heart J **147**: 540-544, 2004.

[15] el-Sherif N, Caref EB, Yin H, Restivo M: The electrophysiological mechanism of ventricular arrhythmias in the long QT syndrome. Tridimensional mapping of activation and recovery patterns. Circ Res **79**: 474-492, 1996.

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