

Images in  
Cardiovascular Medicine



# Congenital Long QT Syndrome Type 2 with Symptomatic 2:1 Atrioventricular Block and Ventricular Arrhythmia in a Preterm Baby Who Presented with Fetal Ventricular Tachycardia and Hydrops

Ju Ae Shin, MD<sup>1</sup>, Yeon U Choi, MD<sup>1</sup>, Kyung Min Kim, MD<sup>2</sup>, Ji Hong Yoon , MD<sup>1</sup>, Jae Young Lee , MD, PhD<sup>1</sup>

<sup>1</sup>Department of Pediatrics, Seoul St. Mary's Hospital, College of Medicine, The Catholic University of Korea, Seoul, Korea

<sup>2</sup>Department of Pediatrics, Incheon St. Mary's Hospital, College of Medicine, The Catholic University of Korea, Seoul, Korea



Received: Jun 8, 2021

Accepted: Aug 4, 2021

## Correspondence to

Jae Young Lee, MD, PhD


Division of Cardiology, Department of Pediatrics, Seoul St. Mary's Hospital, College of Medicine, The Catholic University of Korea, 222, Banpo-daero, Seocho-gu, Seoul 06591 Korea.

E-mail: jaeyounglee@catholic.ac.kr


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## ORCID iDs

Ji Hong Yoon 

<https://orcid.org/0000-0001-8498-2352>

Jae Young Lee 

<https://orcid.org/0000-0003-3347-0626>

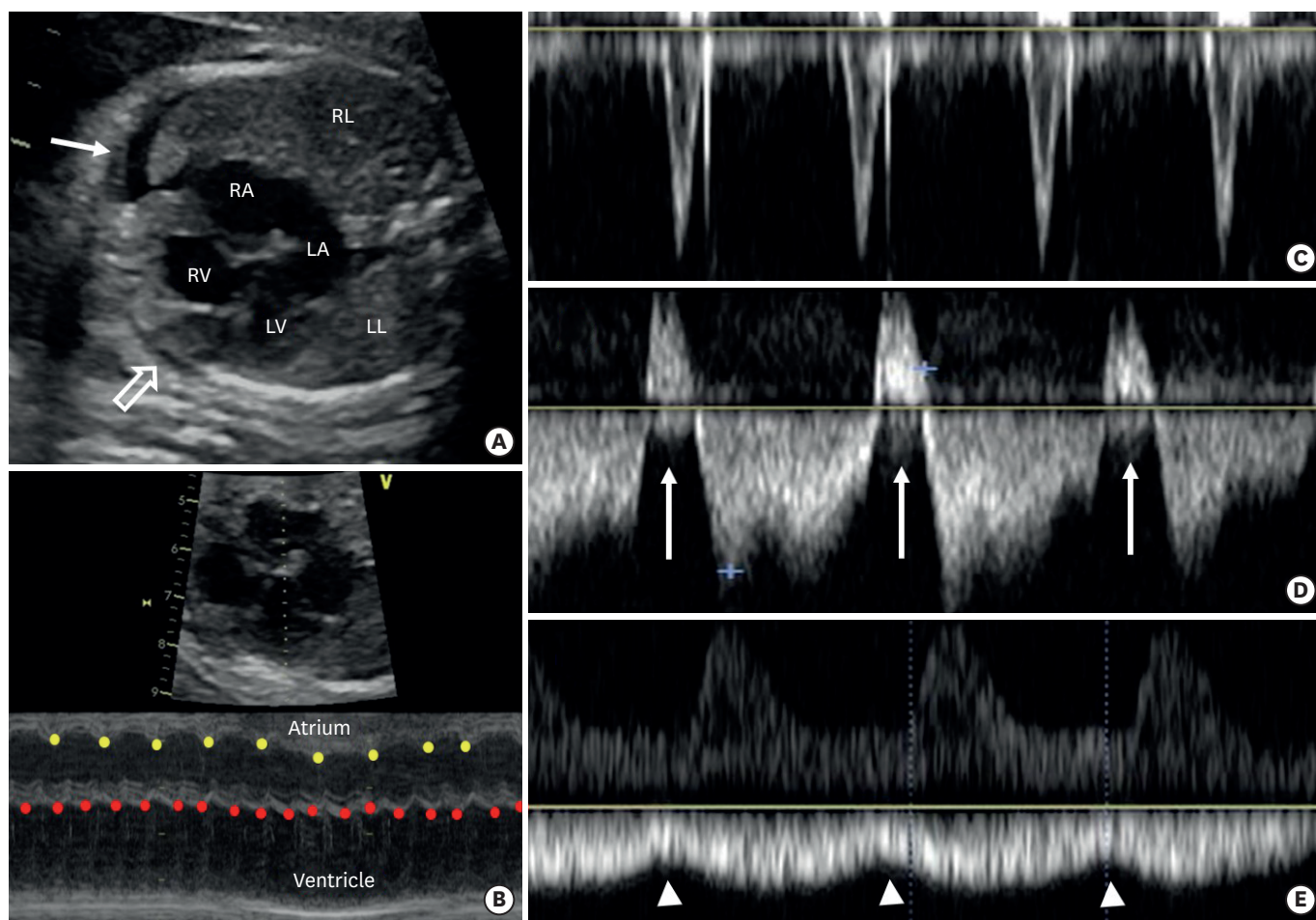
## Funding

The authors received no financial support for the research, authorship, and/or publication of this article.

A baby was born through emergency C-sec delivery at 28<sup>+3</sup> weeks of gestation, with a birth weight of 1.3 kg, due to fetal ventricular tachycardia, severe ventricular dysfunction, and fetal hydrops (**Figure 1**, **Supplementary Videos 1 and 2**).<sup>1)</sup> Electrocardiogram and Holter monitoring showed intermittent 2:1 atrioventricular blocks (AVB) and prolonged QT interval with a circulatory collapse during bradycardia (**Figure 2**). Inotropes including isoproterenol were started, but bradycardia episodes persisted despite decrease in frequency. After a week of unstable clinical conditions, ventricular functions began to improve and the patient began to tolerate bradycardia episodes due to 2:1 AVB. At a month of age, a non-sustained polymorphic ventricular tachycardia (Torsades de Pointes; TdP) developed, 2–3 times a day (**Figure 3**). A beta-blocker was applied without an effect, then oral mexiletine was added, resulting in a gradual decrease of QT interval and the AVB episodes, and eventually TdP disappeared (**Figure 4**). The patient was discharged home at 87 days after birth, taking propranolol and mexiletine. Currently, the patient is 16 months old and in normal growth without any cardiovascular event. The last electrocardiogram showed 1:1 AV conduction with much decreased QTc (470 ms) (**Figures 4 and 5**). Next-generation sequencing revealed a *KCNH2* gene mutation and long QT syndrome (LQTS) type 2 was confirmed.

Ventricular arrhythmia and/or 2:1 AVB associated with LQTS may cause fetal hydrops and intrauterine fetal demise, have been reported to have high mortality rate during infancy, and frequently need device therapy.<sup>2)3)</sup> Mexiletine may be a good alternative choice for these patients.<sup>4)5)</sup>

The Institutional Review Board of Seoul St. Mary's Hospital approved this study and the informed consent was waived (IRB number: KC21ZISI0437).



**Figure 1.** Fetal echocardiography images. (A) A transverse scanning with a four-chamber view shows enlarged right atrium, dilated and hypertrophied right ventricle, small amount of pericardial effusion (open arrow), and pleural effusion (closed arrow). (B) An M-mode image shows a ventricular tachycardia (red dots) with a heart rate of 215 beats/min, instead atrial rate (yellow dots) is 120 beats/min. (C) Tricuspid valve inflow Doppler shows a sharp monophasic filling pattern. (D) A ductus venosus flow Doppler shows reverse flows during atrial contractions (arrows). (E) An umbilical venous flow Doppler shows notches during atrial contractions (arrow heads). All of the above findings suggest severe fetal cardiovascular dysfunction and poor general conditions, which demand immediate treatment.<sup>9)</sup> Image C, D and E were obtained during the time without ventricular tachycardia. LA = left atrium; LL = left lung; LV = left ventricle; RA = right atrium; RL = right lung; RV = right ventricle.

**Conflict of Interest**

The authors have no financial conflicts of interest.

**Presentation**

The case has been presented at the 64<sup>th</sup> Annual Scientific Meeting of the Korean Society of Cardiology in 2020.

**Data Sharing Statement**

The data generated in this study is available from the corresponding author upon reasonable request.

**Author Contributions**

Conceptualization: Lee JY; Methodology: Shin JA, Kim KM, Yoon JH; Supervision: Lee JY; Writing - original draft: Shin JA, Lee JY; Writing - review & editing: Choi YU, Kim KM, Yoon JH.

**SUPPLEMENTARY MATERIALS**

**Supplementary Video 1**

Fetal echocardiography at 28<sup>+3</sup> weeks of gestation showing vary rapid ventricular contractions with a dilated and hypertrophied right ventricle, dilated right atrium, and decreased both ventricular contractions.

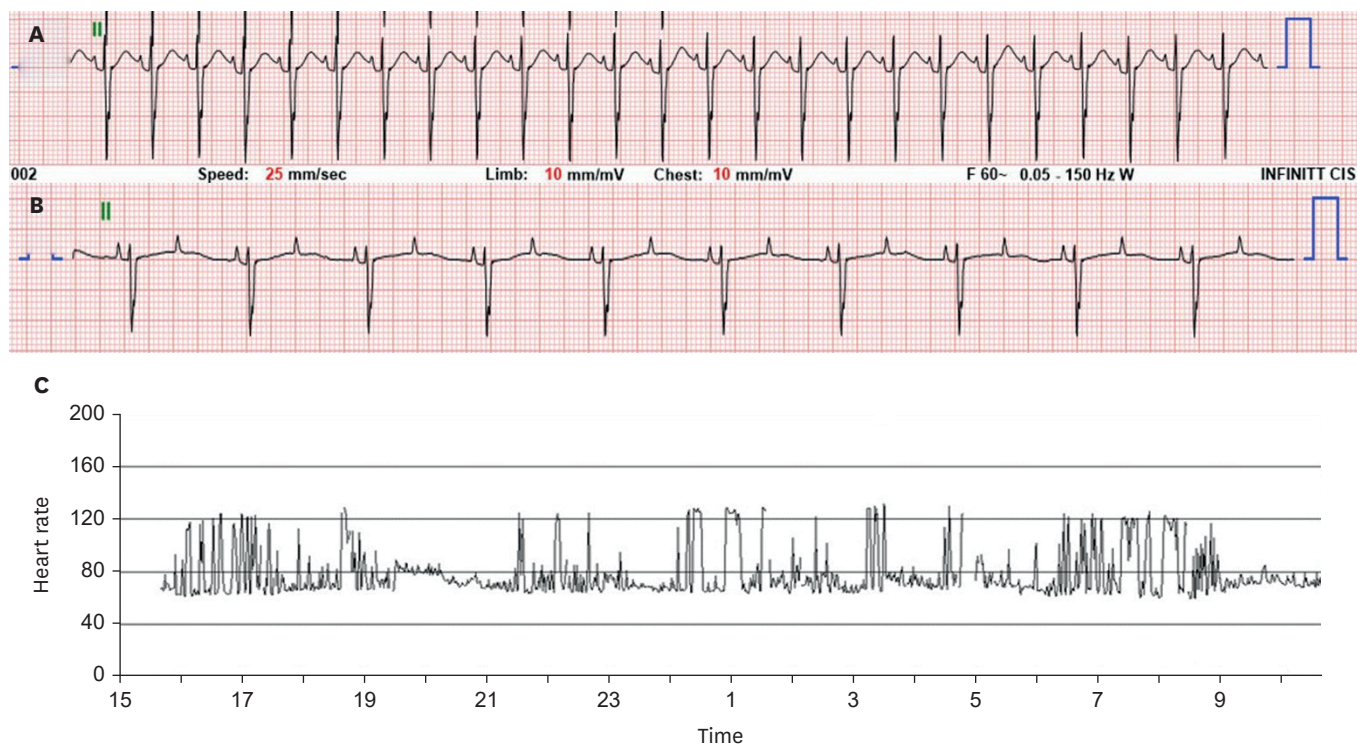
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**Supplementary Video 2**

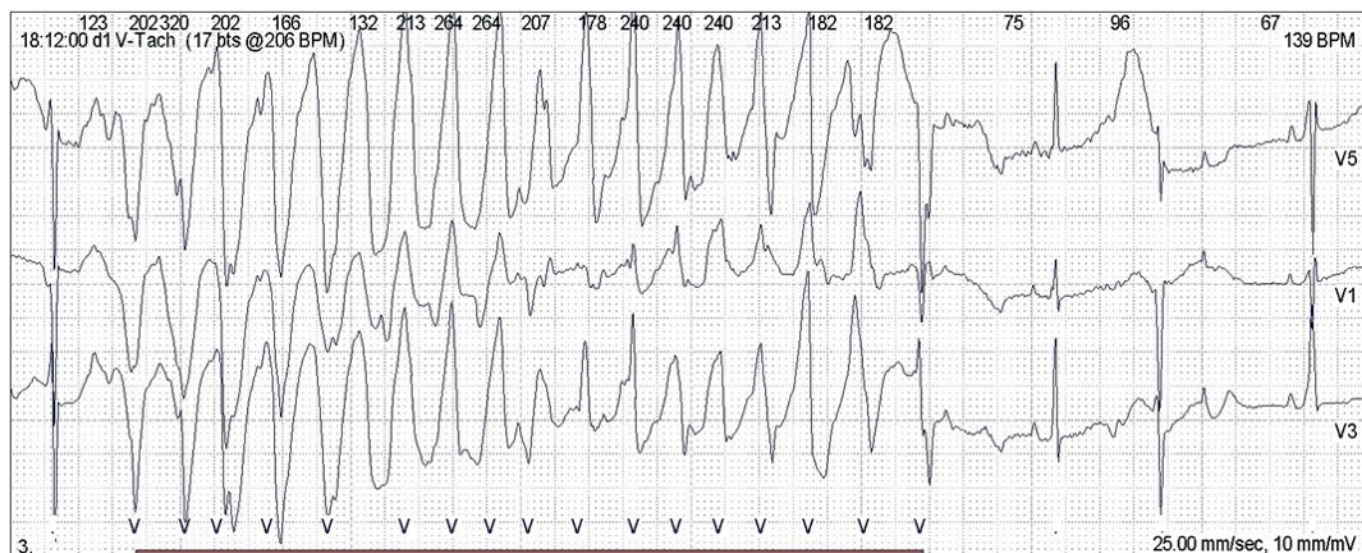
Fetal echocardiography at 28<sup>+3</sup> weeks of gestation. A color flow mapping showed a pericardial effusion, inadequate tricuspid valve inflow, tricuspid regurgitation, and very rapid ventricular rate with decreased contractions.

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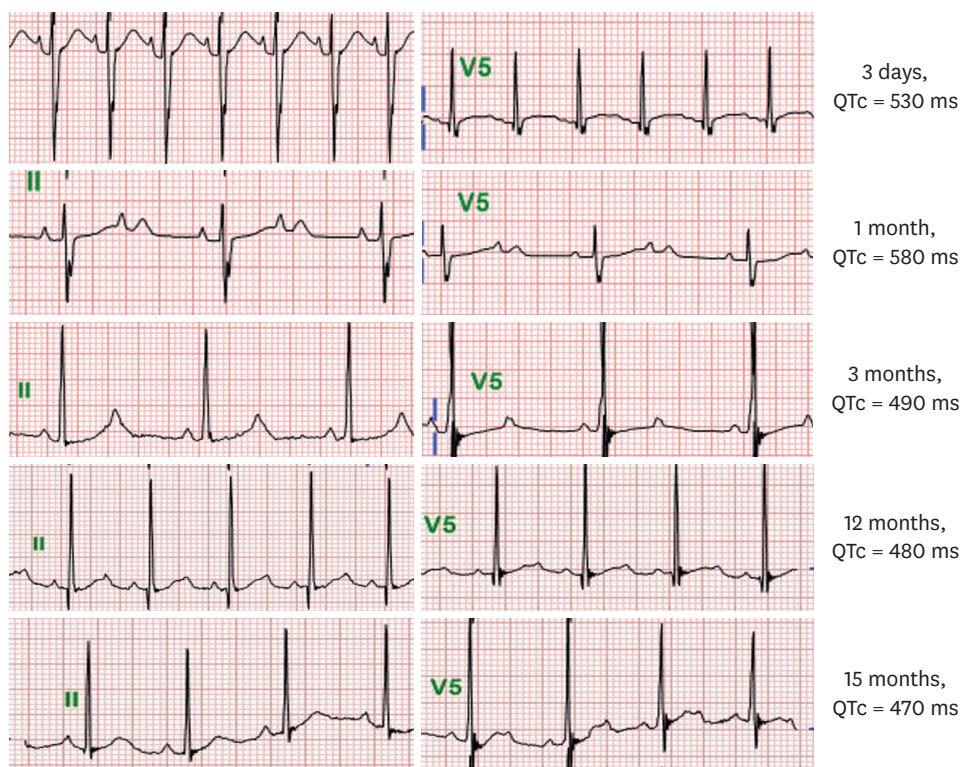


**Figure 2.** Electrocardiograms after birth showing a sinus rhythm with a heart rate of 166 beats/min and prolonged QT interval (QTc = 530 ms) (A) and frequent 2:1 atrioventricular blocks with a heart rate of 50–60 beats/min (B). Severe sudden heart rate fluctuations (50–130 beats/min) were observed on Holter monitoring (C).

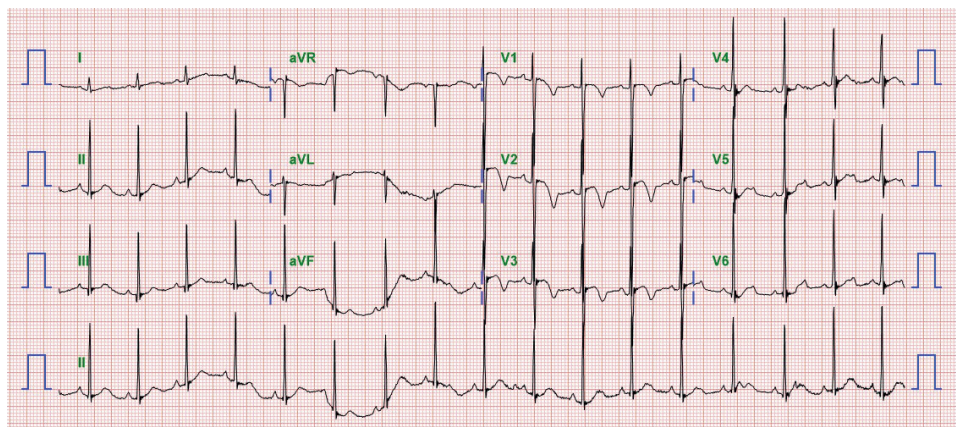


**Figure 3.** Electrocardiogram at 1 month of age, showing a polymorphic ventricular tachycardia (Torsades de pointes), which occurred 2–3 times a day.





**Figure 4.** Serial changes of electrocardiogram (lead II and V5) and corrected QT intervals (QTc) over time according to the age of the patient. Electrocardiograms at 1 and 3 months of age showed 2:1 atrioventricular blocks and thereafter, normal sinus rhythms.



**Figure 5.** Electrocardiogram at 15 months of age showing a sinus rhythm with a QTc of 470 ms, and notched and asymmetrical T-waves, which are rather typical T-wave morphology in LQT2.



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