

## 특발성 Long QT 증후군 1례

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= Abstract =

## A Case of Idiopathic Long-QT Syndrome(LQTS)

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The idiopathic long-QT syndrome is an infrequent inherited disorder, characterized by prolonged QT interval and by the occurrence of life-threatening tachyarrhythmia, particularly in association with emotional or physical stress. In its characteristic presentation, with obvious QT prolongation and stress induced syncope with torsades de pointes, the diagnosis is straightforward for physicians aware of the disease. But sometimes in cases of borderline QT prolongation and vague symptoms, a correct diagnosis is delayed and frequently misdiagnosed as a seizure disorder. The mortality of untreated symptomatic patients with LQTS exceeds 20% in the year after their first syncopal episode and approaches 50% within 10 years. But this high mortality rate has been significantly reduced by the use of pharmacological or surgical antiadrenergic therapy or both. So early detection of the disease and antiadrenergic treatment are important. There are two mechanisms to explain LQTS. First there was an imbalance in the sympathetic innervation to the heart. Second, there was intrinsic gene abnormality in the mechanisms responsible for cardiac repolarization.

We report one case of LQTS in a 24 years old female patient who had been suffering from stress related syncope. Her initial EKG had prolonged QT interval(588msec) and increased QT interval dispersion(200msec). In the Holter monitor, long and short cycle sequence induced(pause-dependent) torsades de pointes was detected. She has been treated by  $\beta$ -blocker and has maintained her condition without recurrence of the symptoms.

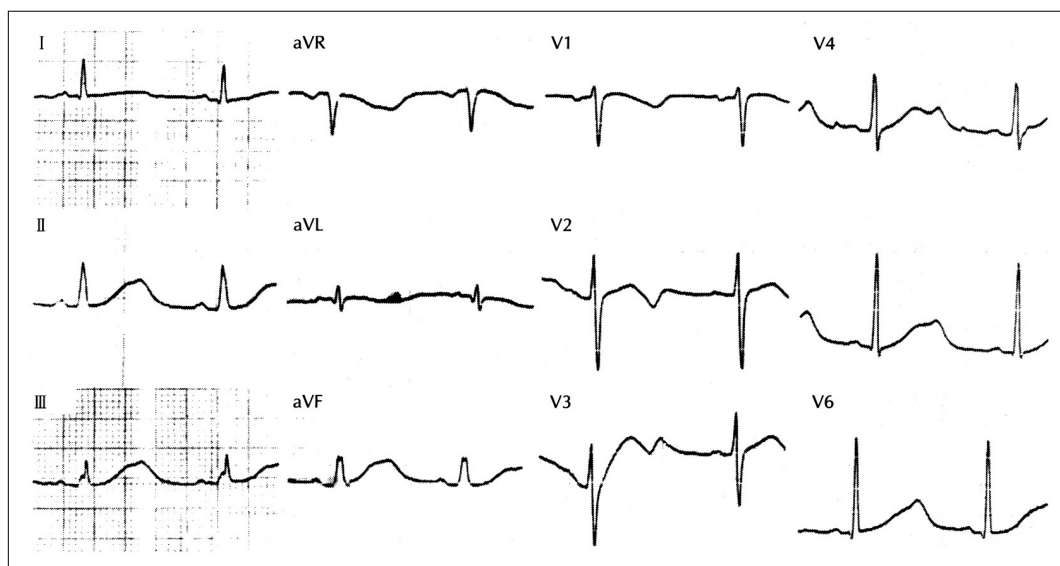
**KEY WORDS** : Long QT syndrome · Torsades de pointes ·  $\beta$ -blocker.

서 론

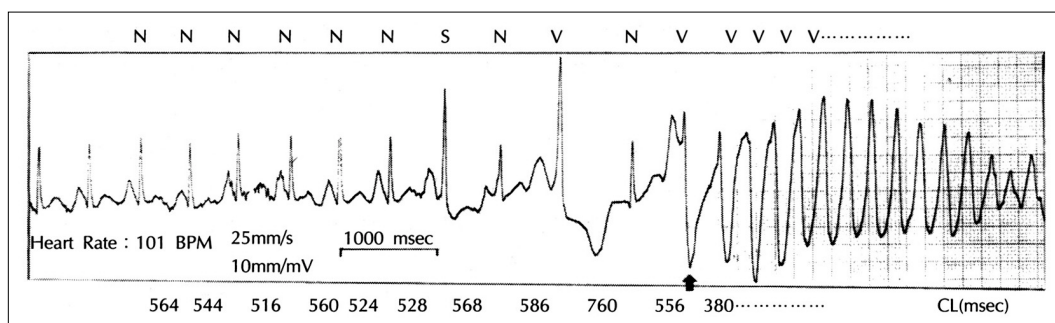
1) .

Long QT ( LQTS) 가 가 , ,  
QT

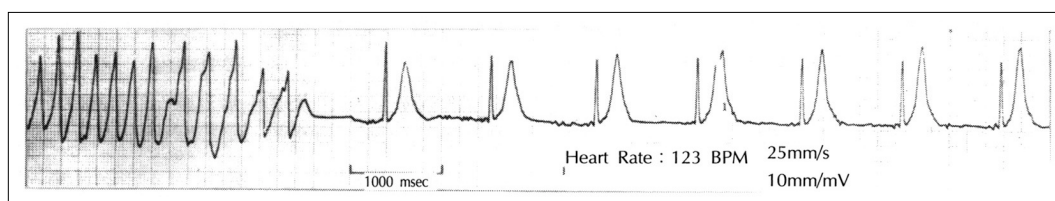
가 , 가 , 24  
 가 20%가  
 가 2)  
 가 3)  
 5% : 5  
 4)  
 QT  
 가 : 가  
 , 3  
 QT 24  
 Torsades de pointes Long QT : 가  
 1 : 130/90mmHg,  
 74 , 20 / , 36.8  
 증 례  
 : , 24 ,  
 :  
 : 2 2 3 7,000/mm<sup>3</sup>, 12.0g/dl, 36%,  
 287,000/mm<sup>3</sup> FBS  
 2 4 85mg/dl, 146mg/dl, 75mg/dl,  
 2 3 HDL 48.2mg/dl ,



**Fig. 1.** An electrocardiogram(EKG) on first visiting day.

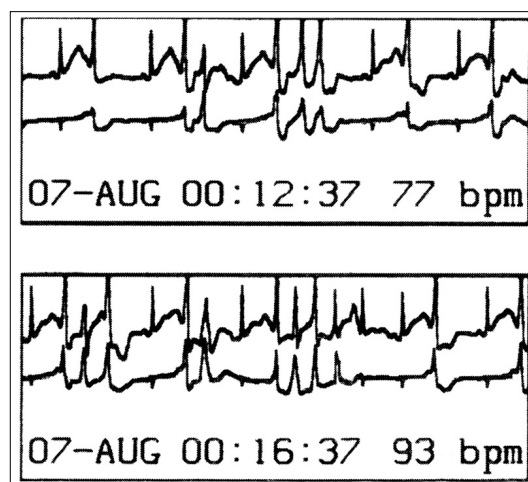


**Fig. 2.** Cascade phenomenon, where a premature atrial beat initiated a short-long phenomenon leading to the onset of a first premature ventricular beats, whose compensatory pause triggered the onset of torsade de pointes. Note the major changes in T wave configuration in the beat after the postextrasystolic pause. Paper speed 25mm/s. N(S.V)=normal(supraventricular, ventricular) beat.



**Fig. 3-1.** R-R interval prolongation and QT interval shortness in time of torsade de pointes spontaneously disappear.

X - ( ) .  
 (Fig. 1) 67 /min,  
 QT/QTc 588/603msec, QT dispersion(  
 12 가 QT 가  
 QT ) 200msec U 가  
 24 3  
 Torsades de pointes가  
 Long-short cycle sequence cascade phen-  
 omenon Torsades de pointes  
 (Fig. 2), Torsades de pointes가  
 R-R QT  
 (Fig. 3 - 1).



**Fig. 3-2.** Salvos of 2 to 4 consecutive beats.

(Salvos of 2 to 4 consecutive beats)(Fig. 3 - 2)  
 (Bigeminy of premature ventricular  
 beats)(Fig. 3 - 3)

Torsade de points가

: (pro - QTc dispersion (Table 1), QT/  
 ( 6 ) QT/QTc 50  
 pranolol 160mg , QT/QTc dispersion 120/1  
 4 4/496msec

10msec  
xolol 20mg PO qd)

(Beta -

가, 5

QT/QTc

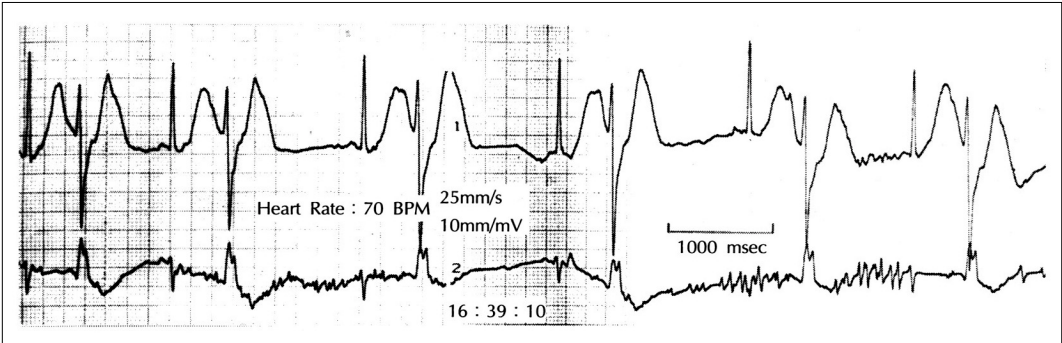


Fig. 3-3. Bigeminy of premature ventricular beats.

Table 1. Changes of QT/QTc and QT/QTc dispersion during admission period

Hospital day	H · R (beats/min)	QT (msec)	QTc (msec)	QT disp. (msec)	QTc disp. (msec)
#1	76	580	610	230	250
#3	58	512	508	200	180
#5	57	532	527	160	180
#6 (Discharging day)	56	504	496	100	95

H · R : Heart rate, QT : QT interval, QTc :  $QT/\sqrt{RR}$  (Bazett's formula)  
QT disp. : Dispersion of QT (QTmax-QTmin) QTc disp. : Dispersion of QTc (QTcmax-QTcmin)

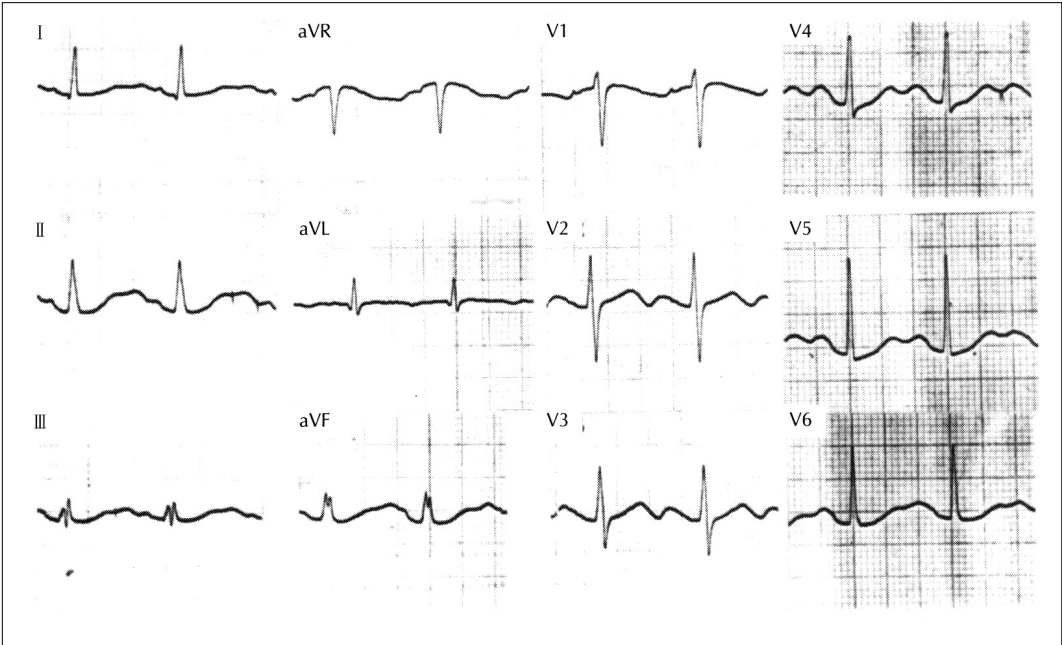


Fig. 4. An electrocardiogram (ECG) after 5.5 months later.

440/493msec QT/QTc dispersion 180/  
200msec (Fig. 4).

가  
가 LQTS QT/QTc  
456/486 msec, QT/QTc dispersion가 100/10  
5msec  
3

고 안

Long QT

QT

T U

Jervell - Lange - Nielsen

5)

Romano - Ward

6)

가

. Schwartz

0.25%

7)

0.49%

Jervell - Lange - Nielsen

Romano - Ward

LQTS 가

**Table 2.** 1993 LQTS diagnostic criteria

	Points
ECG findings*	
A. QTc †	
480msec <sup>1/2</sup>	3
460 - 470msec <sup>1/2</sup>	2
450msec <sup>1/2</sup> (in males)	2
B. Torsades de pointes ‡	1
C. T-Wave alternans	1
D. Notched T wave in three leads	0.5
E. Low heart rate for age	
Clinical history	
A. Syncope ‡	
With stress	2
Without stress	1
B. Congenital deafness	0.5
Family history	
A. Family members with definite LQTS #	1
B. Unexplained sudden cardiac death below age 30 among immediate family members	0.5

LQTS, long QT syndrome

\*In the absence of medications or disorders known to affect these electrocardiographic features

† QTc calculated by Bazett's formula, where QTc=QT/h

‡ Mutually exclusive

Resting heart rate below the second percentile for age

The same family member cannot be counted in A and B

# Definite LQTS is defined by an LQTS score 4

Scoring : 1 point, low probability of LQTS ; 2 to 3 points, intermediate probability of LQTS ; 4point, high probability of LQTS

가

1993 LQTS

8)

가

3가

0

9

가

(Probability)

(Table 2).

. Schwartz

QTc가 440msec

가

, LQTS 가

1985 LQTS

QTc가 0.46sec

가

0.41 0.45sec

가

5 10%

LQTS

5%

QT

(heterog -

eneity)

QT dispersion<sup>9)</sup>(QTmax - Qtmin)

LQTS

QT dispersion

가 480msec

470msec

100% 가

가

QT

QTc

가 440msec

QTc 가 LQTS

가

45msec LQTS 135msec LQT1 LQT2 LQT4  
. Priori <sup>10)</sup>  
QT dispersion LQTS Jervell - Lange - Nielsen  
LQTS 가 QT dispersion LQT3 sodium channel inactivation defect가 QT torsade de pointes  
가 sodium channel blocker mexiletine  
. LQT2 potassium channel gene defect QT  
QT dispersion 100 msec torsade de pointes potassium channel opener가  
QT 가 . LQT1 potassium channel LQTS 50%  
가 . T <sup>11)</sup> KVLQ1 가  
가 LQTS 가  
U LQTS T - wave alternans U - wave alternans LQTS  
LQTS Torsades de pointes 1)  
가가 early afterdepolarizations(EADs)  
가 (triggered activity) <sup>18)</sup>  
<sup>12)</sup> 가 2)  
( (M - cell layer)  
) , reentry (unidirectional block)  
ion channel , M - cell reentry  
가 <sup>13)</sup> ion channel 가 reentrant excitation torsades de pointes가 <sup>19)</sup>  
(mutation)가 LQTS  
LQTS 90% dispersion  
가 ion channel 가 가 EADs  
( post - extrasystolic pauses)가  
가 dispersion 가  
1) 11 LQT1 : EADs  
KVLQT1 locus <sup>14)</sup>, 2) 7 LQT2 : torsades de pointes  
HERG locus <sup>15)</sup>, 3) 3 LQT3 :  
SCN5A locus <sup>16)</sup>, 4) torsades de pointes  
4 LQT4 locus <sup>17)</sup>가

## References

- 1) Schwartz PJ, Periti M, Malliani A : *The long QT syndrome*. *Am Heart J* 89 : 378, 1975
- 2) Schwartz PJ : *Idiopathic long QT syndrome : Progress and questions*. *Am Heart J* 109 : 399, 1985

- 664 -

- long QT syndrome to chromosome 4q25-27. Am J Hum Genet* 57 : 1114, 1995
- 18) Brachmann J, Scherlag BJ, Rosenshtraukh LV, Lazzara R : *Bradycardia-dependent triggered activity : Relevance to drug-induced multiform ventricular tachycardia. Circulation* 68 : 846, 1983
  - 19) Pertsov AM, Davidenko JM, Salomonsz R, Baxter WT, Jalife J : *Spiral waves of excitation underlie reentrant activity in isolated cardiac muscle. Circ Res* 72 : 631, 1993
  - 20) Viskin S, Alla SR, Barron HV, Heller K, Saxon L, Kitzis I, van Hare GF, Wong MJ, Lesh MD, Scheinman MM : *Mode of onset of torsade de pointes in congenital long QT syndrome. J Am Coll Cardiol* 28 : 1262, 1996
  - 21) Schwartz PJ, Locati EH, Moss AJ, Crampton RS, Trazzi R, Ruberti U : *Left cardiac sympathetic denervation in the therapy of the congenital long QT syndrome : A worldwide report. Circulation* 84 : 503, 1991
  - 22) 최기준 · 이철환 · 김재중 · 김유호 : *Long QT 증후군 환자에서의 삽입형 심실제세동기 치료. 순환기* 26 : 1198, 1996