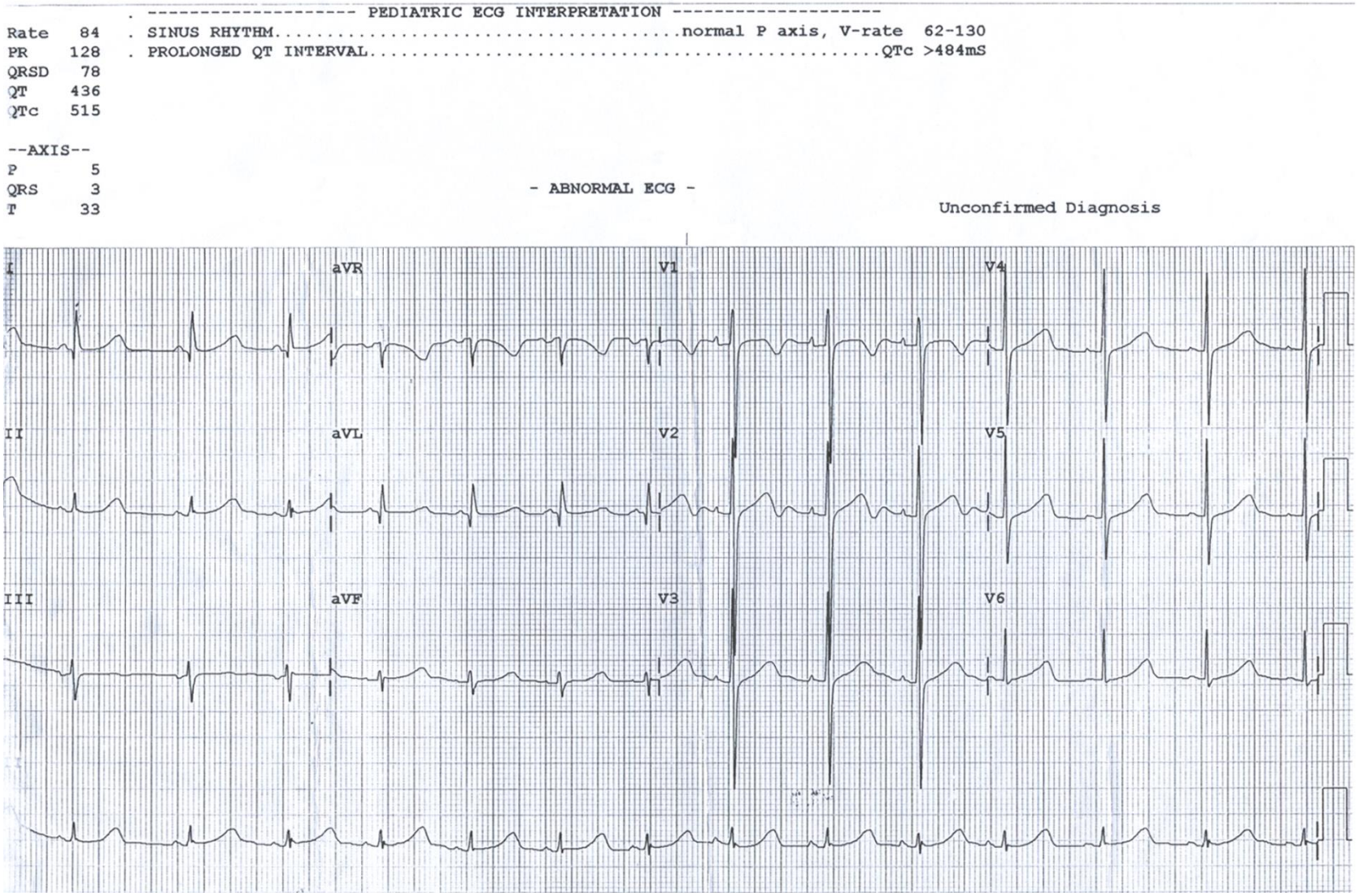
Left Cardiac Sympathetic Denervation

경북대 조용근

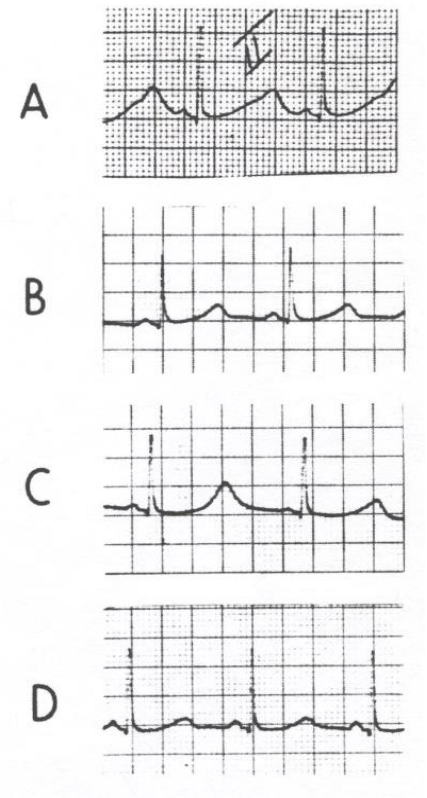
축구를 하다 실신하는 9살 남아



Unilateral cervicothoracic sympathetic ganglionectomy for the treatment of LQTS

Moss and McDonald, NEJM 1971;285:903-4, Lowengrub, Prog Cardiovasc Dis 2015;58:221-6

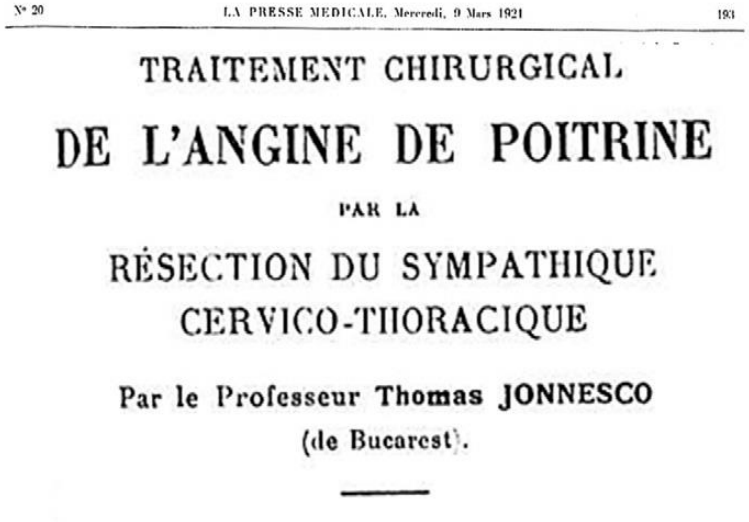
- An adult white female experienced her first syncopal episode at age 39 in 1968, and she had four additional fainting episodes in the next 18 months, with most episodes occurring in situations in which she was acutely excited or emotionally stressed.
- The patient had a syncopal spell when a blood sample was being withdrawn from a vein with pain and discomfort at the vein site. Polymorphic VT at a rate of >200 beats/min was documented.
- After ganglionectomy the patient has continued to be followed clinically by Dr. Moss since 1970, and presently (2015) she is alive and well at age 85 without recurrent syncope.



- In A, one day after the patient's first syncopal episode (QT = 0.64 s).
- In B, 83 minutes after local left stellate-ganglion block (QT = 0.46 s).
- In C, 40 minutes after local right stellate ganglion bloc (QT = 0.72 s).
- In D, six months after left cervicothoracic sympathetic ganglionectomy (QT = 0.44 s).

History of Sympathectomy

Odero, Heart Rhythm 2010;7:1161-5



N° 20 LA PRESSE MEDICALE, Mercredi, 9 Mars 1921 193

TRAITEMENT CHIRURGICAL

DE L'ANGINE DE POITRINE

PAR LA

RÉSECTION DU SYMPATHIQUE CERVICO-THORACIQUE

Par le Professeur Thomas JONNESCO

(de Bucarest).

Jonnesco, Prese Med 1921;20:193-4
from Schwartz, Int J Cardiol 2017;237:25-8

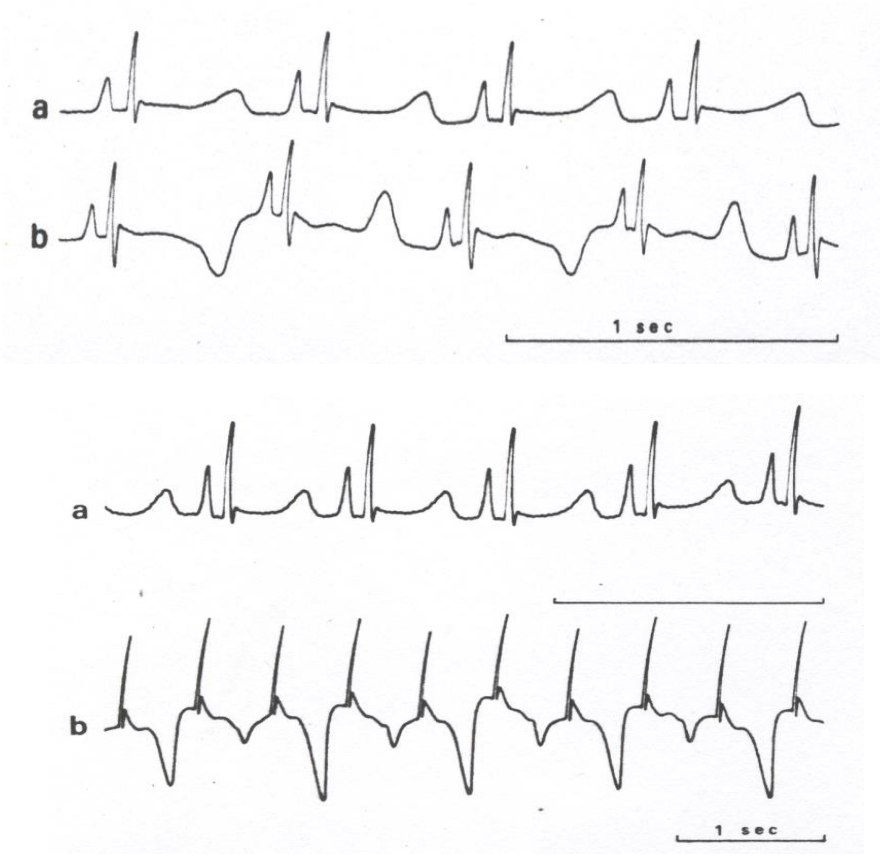
- Jonnesco, in 1916, for the first time performed unilateral section of the left stellate ganglion in a patient affected by incapacitating angina with cardiac arrhythmias. After surgery, the arrhythmias as well as the attacks of angina disappeared.
- In 1921, Jonnesco confirmed in a larger number of patients that stellate ganglionectomy represented a mainstay in the treatment of angina.
- In 1928, Leriche and Fontaine stated that the left stellate ganglion had a central role in the reflexes that initiate angina and that sympathetic nerves had a vasoconstrictive effect on the coronary arteries.
- A large series of studies showed that sympathectomy were highly successful in preventing anginal attacks and improving performance.
- With the advent of β-blockers, sympathectomy became superseded despite its clear efficacy.

Electrical alternation of the T-wave

Clinical and experimental evidence of its relationship with the sympathetic nervous system and with the long Q-T syndrome

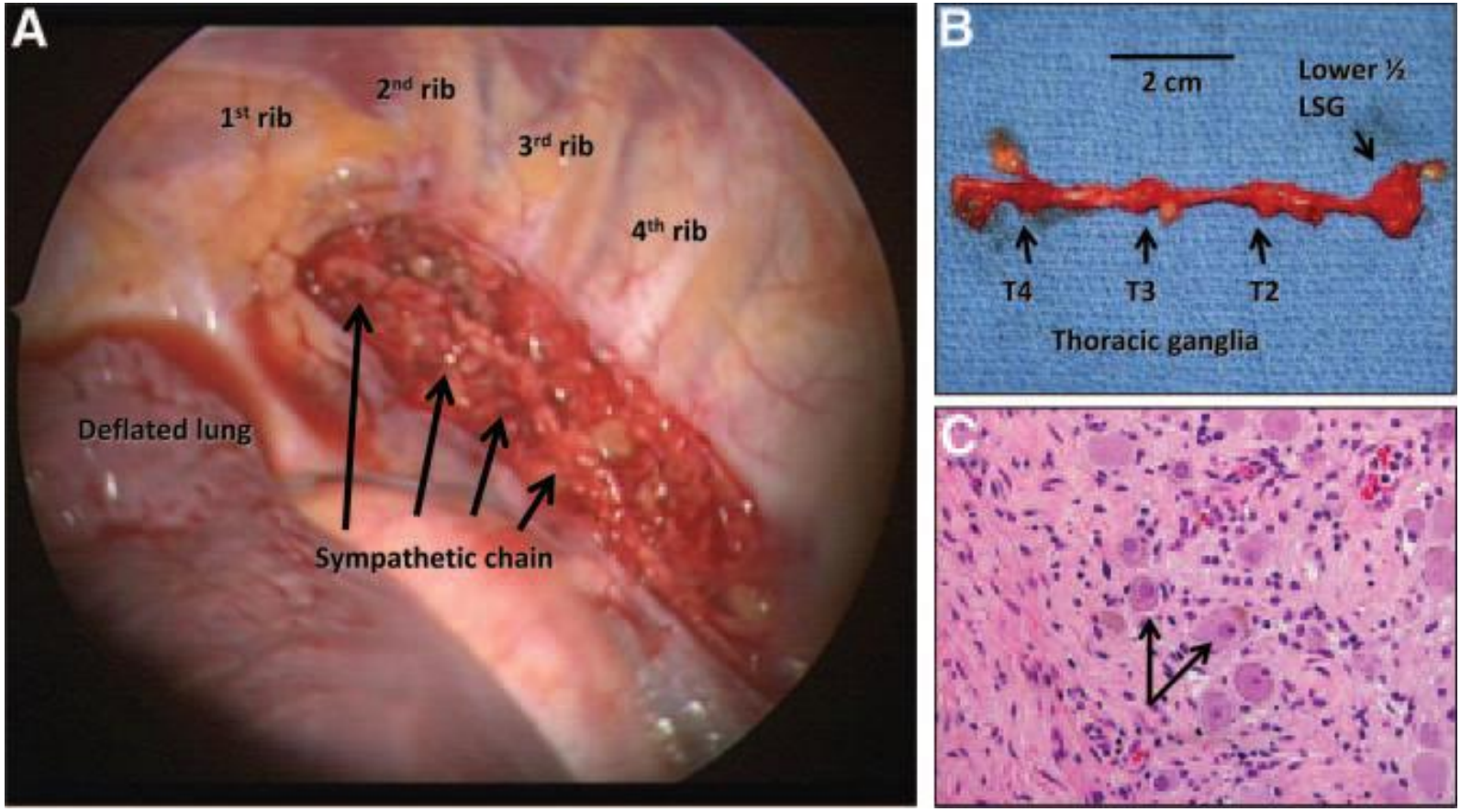
Schwartz, Am Heart J 1975;89:45-50

- The experimental reproduction of both QT prolongation and macroscopic TWA in cats by stimulation of the left stellate ganglion, which highlighted the critical role of the left cardiac sympathetic nerves in triggering life-threatening arrhythmias.



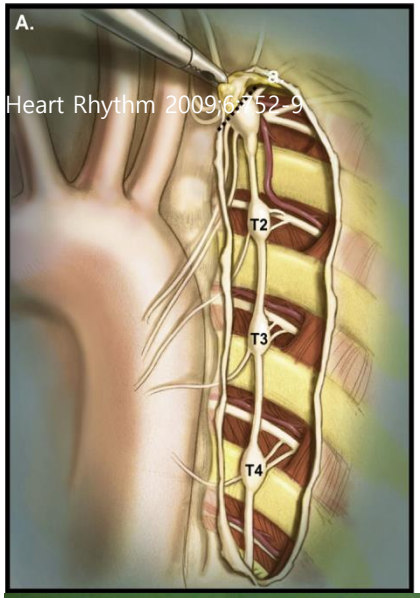
Anatomy and histology of left sympathetic chain

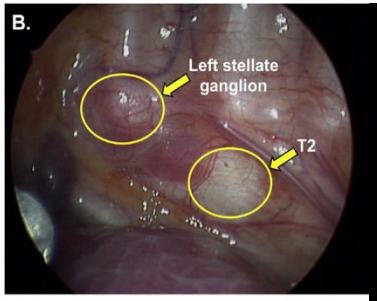
Bourke, Circulation 2010;121:2255-62

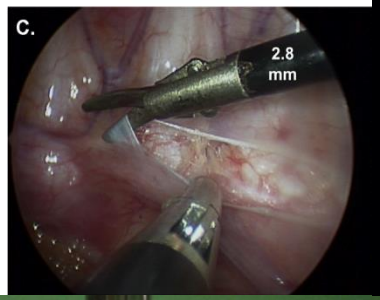


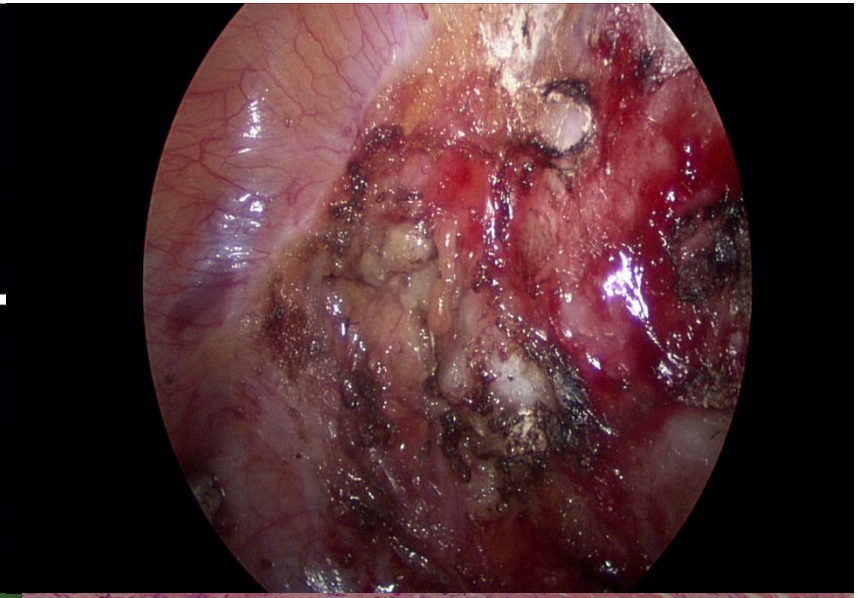
Video-assisted thoracoscopic (VAT) LCSD

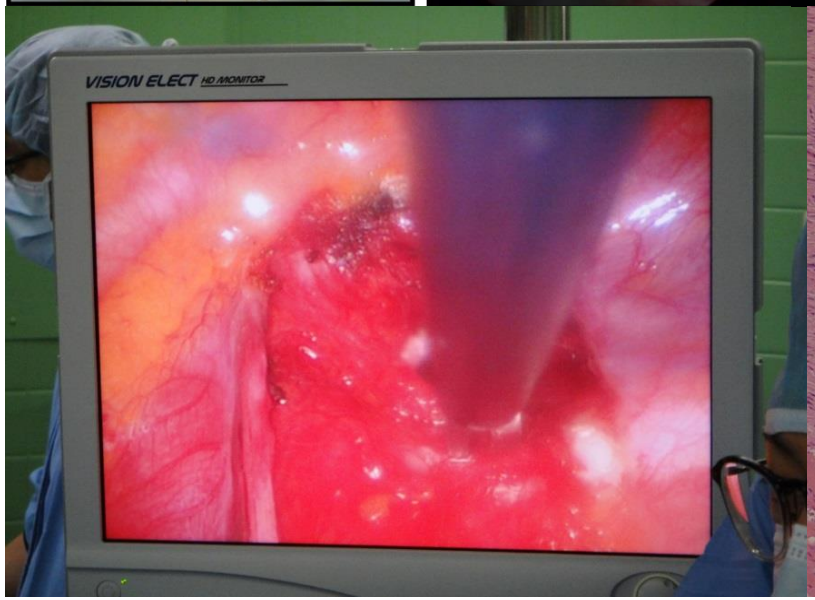
Video-assisted thoracoscopic (VAT) LCSD

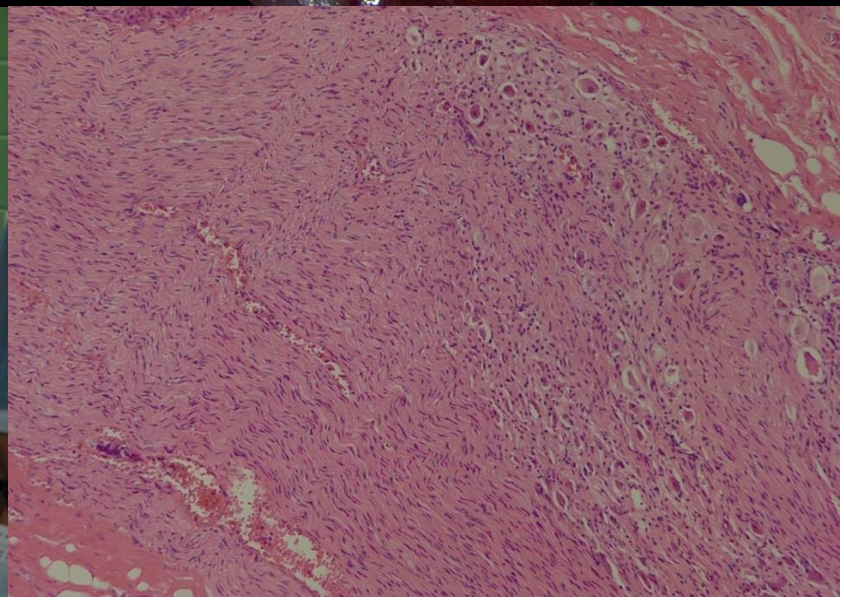






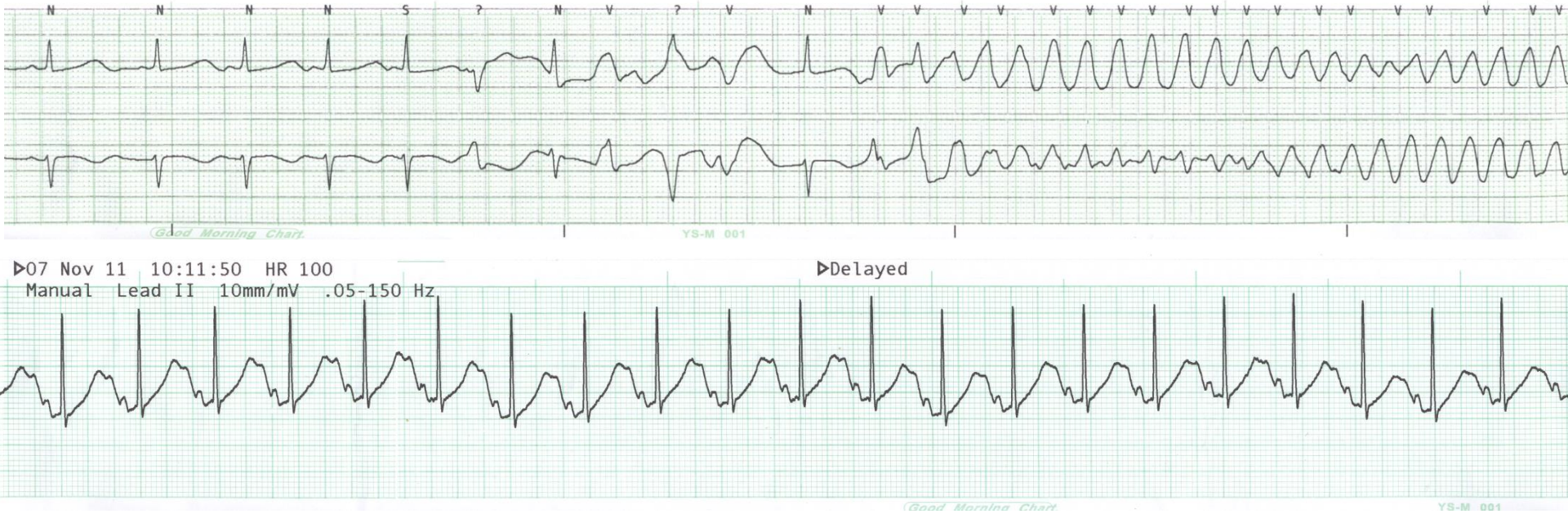






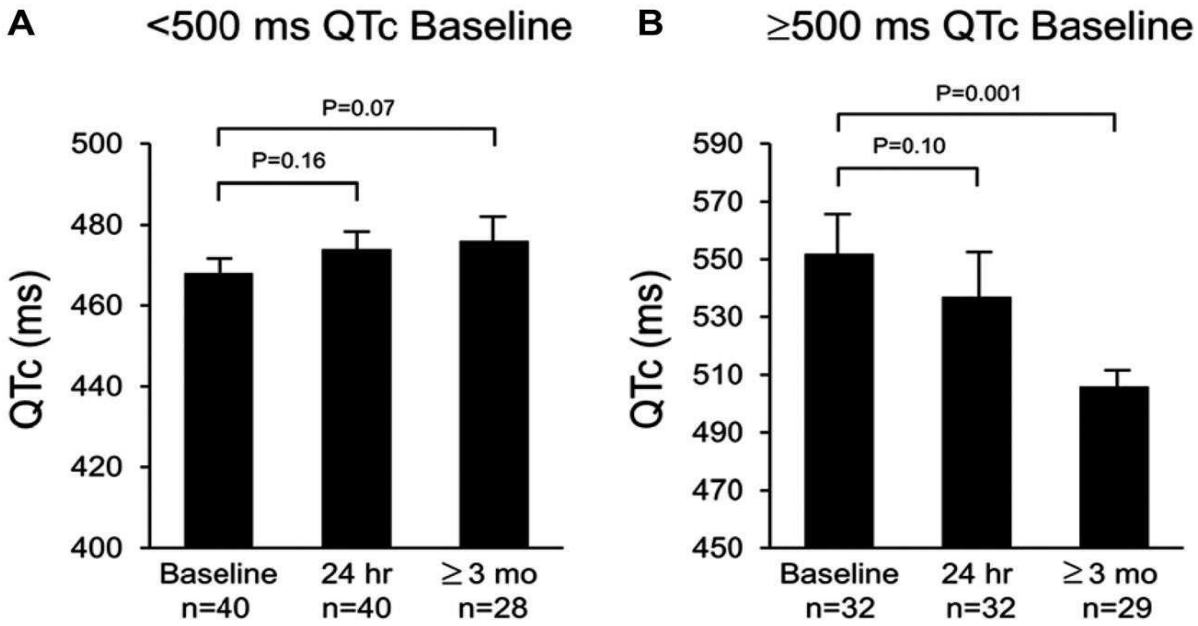
심정지로 내원한 28세 산모

Epinephrine tests: before and after LCSD



Effects on QTc in LQTS patients following LCSD

Desimone, J Cardiovasc Electrophysiol 2015;26:434-9



- 72 patients with LQTS who underwent videoscopic LCSD.
- The majority (74%) had a transient increase in QTc from baseline, with an average maximum increase of 72±30 ms. Resolution within 10 ms of baseline occurred in 57% at 24 hours post-LCSD.
- No patients experienced any arrhythmias related to this transient rise in QTc.
- **The efficacy of LCSD should not be based simply on QTc normalization alone**, but rather on a decrease in arrhythmia-related syncopal events,

Peter J. Schwartz, MD

Head, Center for Cardiac Arrhythmias of Genetic Origin
Director, Cardiovascular Genetics Laboratory, IRCCS Istituto Auxologico Italiano, Milan, Italy

- Schwartz, AJC 1976;37:1034-40. Effect of unilateral cardiac sympathetic denervation on the VF threshold.
- Schwartz, Circ Res 1978;43:939-40. Unilateral stellectomy and dysrhythmias.
- Schwartz, Ann N Y Acad Sci 1984;427:199-221. The rationale and the role of left stellectomy for the prevention of malignant arrhythmias.
- Schwartz, Circulation 1987;76 (I Pt 2):1215-9. SCD. Nonpharmacologic interventions.
- Schwartz, Circulation 1991;84:503-11. LCSD in the therapy of congenital LQTS.
- De Ferrari, J Interv Cardiol 1995;8(6 suppl):776-81. LCSD in the LQTS patients.
- Schwartz, Circulation 2004;109:1826-33. LCSD in the management of high-risk LQTS patients.
- Wilde, NEJM 2008;358:2024-9 LCSD for CPVT.
- Schwartz, Heart Rhythm 2009;6:760-3. Cutting nerves and saving lives.
- Odero, Heart Rhythm 2010;7:1161-5. LCSD for the prevention of life-threatening arrhythmias.
- Schwartz, Nat Rev Cardiol 2014;11:346-53. Cardiac sympathetic denervation to prevent life-threatening arrhythmias.
- De Ferrari, Circulation 2015;131:2158-93. Clinical management of CPVT: The role of LCSD.
  - LCSD is an effective antifibrillatory intervention for patients with CPVT. Whenever syncope occurs despite optimal medical therapy, LCSD could be considered the next step rather than an ICD and could complement ICDs in patients with recurrent shocks.

My approach to the LQTS

Schwartz, Trends Cardiovasc Med 2015;25:376-7

- If I regard the patient as is still at risk despite full-dose β-blockade, I then perform LCSD **without hesitation.**
- This intervention, performed once and for all, makes it more difficult for a heart to fibrillate and still allows for additional therapy.
- I decide on LCSD if the patient has syncope despite β-blockers, when β-blockers are not tolerated, sometimes in the presence of other risk factors such as
  - the occurrence of macroscopic T-wave alternans or
  - a QTc >550 ms, or
  - if the patient has a double mutation.

Michael J. Ackerman, MD, PhD

Professor of Medicine, Pediatrics, and Pharmacology,
Mayo Clinic, Rochester, Minnesota

- Collura, Heart Rhythm 2009;6:752-9. LCSD for the treatment of LQTS and CPVT using video-assisted thoracic surgery.
- Coleman, Circ AE 2012;5:782-8. Videoscopic LCSD for patients with recurrent VF/malignant ventricular arrhythmia syndromes besides congenital LQTS.
- Bos, Circ AE 2013;6:705-11. LCSD in LQTS: Analysis of therapeutic nonresponders.
- De Ferrari, Circulation 2015;131:2158-93. Clinical management of CPVT: The role of LCSD.
- Desimone, J Cardiovasc Electrophysiol 2015;26:434-9. Effects on repolarization using dynamic QT interval monitoring in long-QT patients following LCSD.
- Antinel, Heart Rhythm 2016;13:62-9. Quality of Life after videoscopic LCSD in patients with potentially life threatening cardiac channelopathies/cardiomyopathies.
  - The majority of patients/parents reported that they were very or somewhat satisfied with their surgery (or their child's surgery) and would definitely/probably recommend LCSD to another patient.

My approach to the LQTS

Ackerman, Trends Cardiovasc Med 2015;25:67-9

- Over 1/3 of our LCSDs are performed in patients with unacceptable β-blocker side effects as the primary indication while the 2/3 majority is comprised of patients with a perceived or proven high-risk phenotype.
- The comorbidities associated with LCSD are much less than the ICD-related comorbidities.
- LCSD's anti-fibrillatory effect is greatest in LQT1 followed by LQT2 and LQT3. None of our patients with single LQT1-causative mutations have experienced an LQT1- triggered event since their LCSD. Although most denervated patients remain on β-blocker therapy, 10 LQT1 patients have opted for LCSD monotherapy, with no events thus far.
  - LCSD alone in 18 (3%) patients (Rohatgi, JACC 2017;70:453-62).
- Our present indications for LCSD include a LQTS patient with 1) VF-terminating ICD shocks, 2) β-blocker intolerance, or 3) perceived high risk where combination **β-blocker + LCSD** therapy is used in hopes to avoid or delay an ICD.
- Despite being a referral center for VAT-LCSD, <15% of patients have received LCSD.

APHRS2017

- QTc interval is decreased in 30% of patients with LQTS after sympathectomy.
- Smpathectomy, not touching a stellate ganglion, is a Sham surgery.

저희는

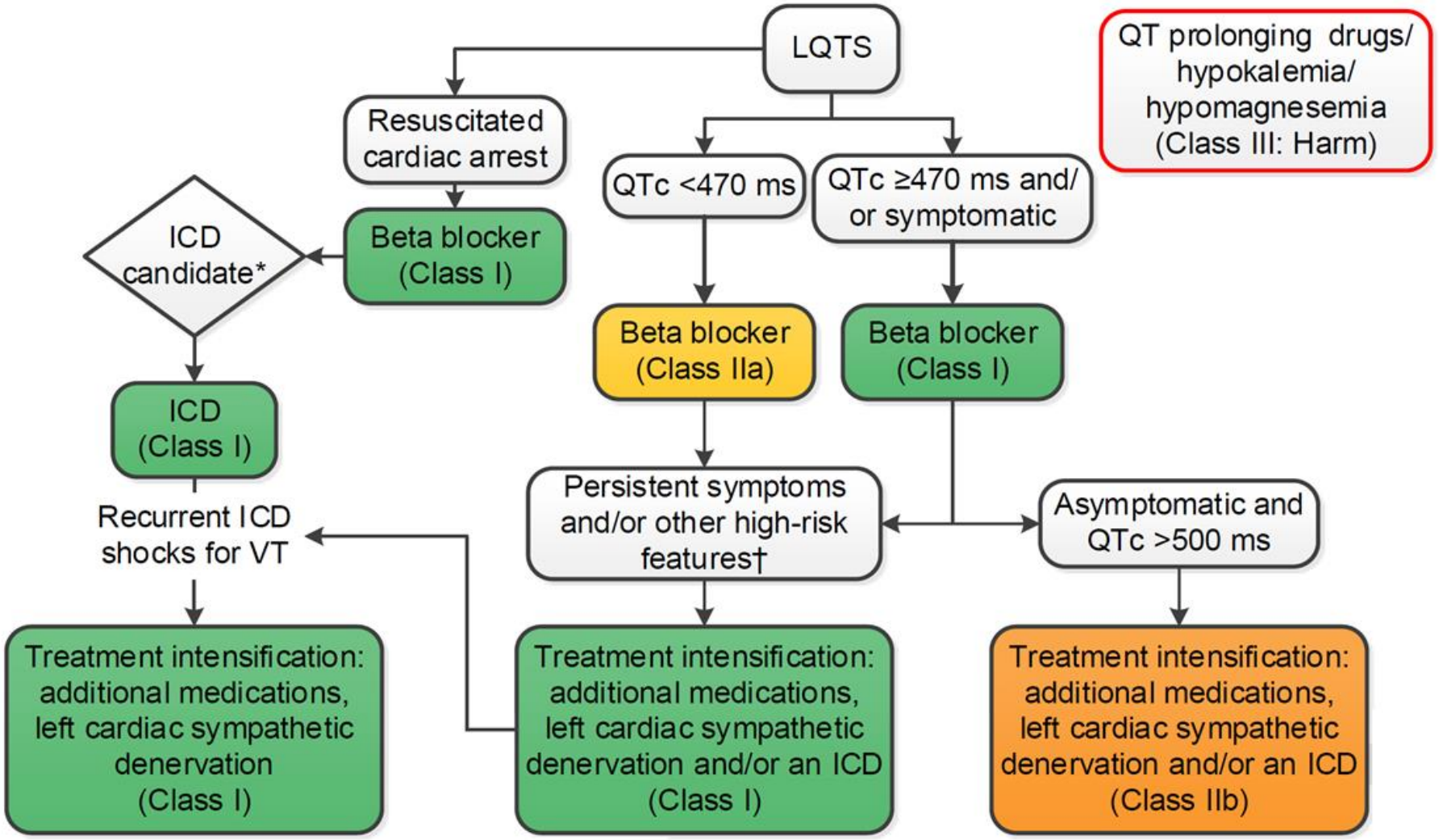
- At CCU
  - Pre-LCSD epinephrine test
  - LCSD: VAT
    - Lower half of stellate ganglion, T2, T3, and T4.
    - Operation time ≒ 1 hr
  - Post-LCSD epinephrine test
  - Removal of chest tube
  - Stays ≒ 3 nights
- Complications
  - No sweating in the left side of face and arm
  - Compensatory sweating in the right side of face and arm
  - Atypical chest pain
  - Horner syndrome: only in 1, resolved spontaneously

2017 AHA/ACC/HRS guideline for management of patients with ventricular arrhythmias and the prevention of sudden cardiac death

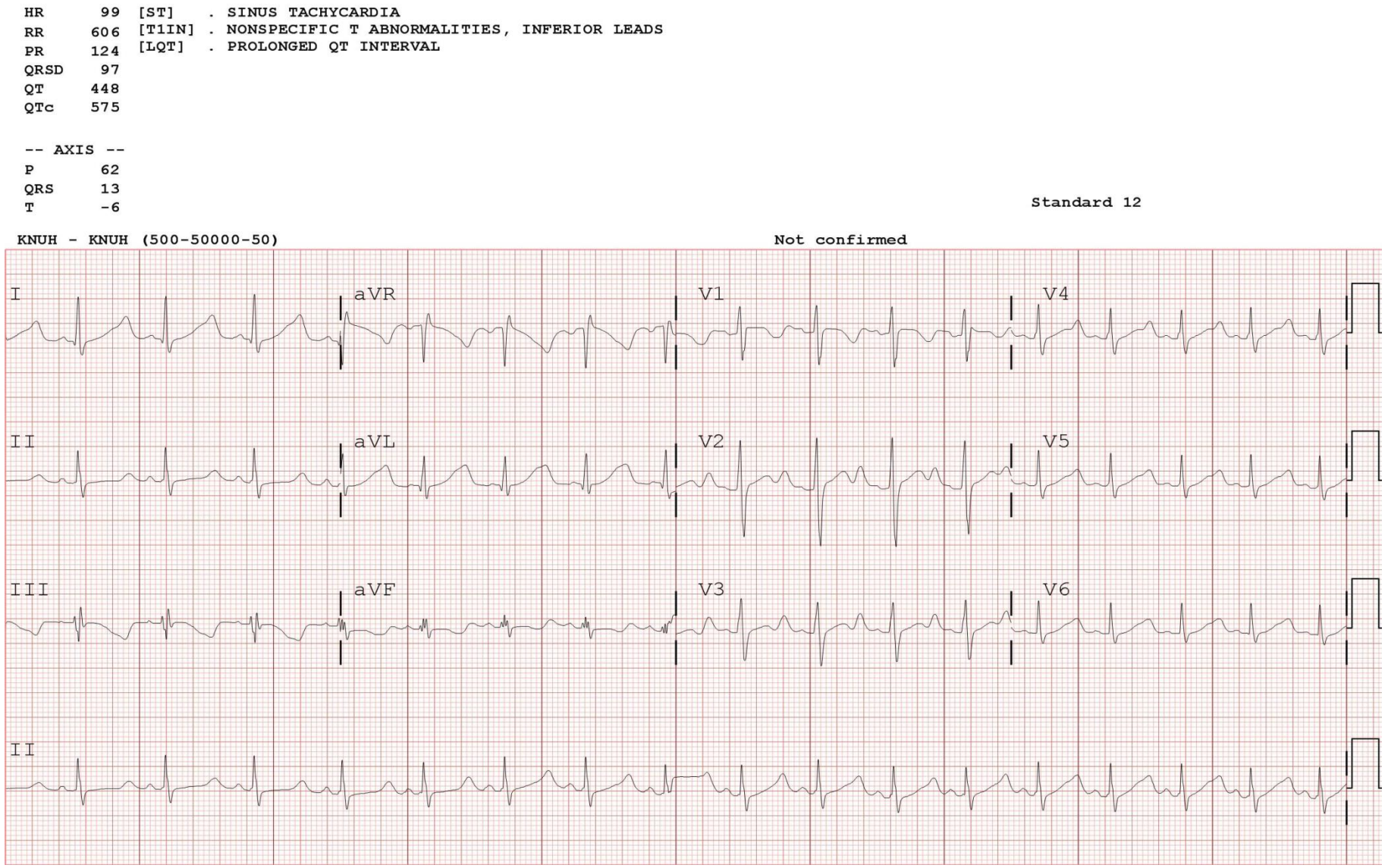
Recommendations for Long QT Syndrome References that support the recommendations are summarized in Online Data Supplement 40.		
COR	**LOE**	**Recommendations**
I	B-NR	1. In patients with long QT syndrome with a resting QTc greater than 470 ms, a beta blocker is recommended (1-5).
I	B-NR	2. In high-risk patients with symptomatic long QT syndrome in whom a beta blocker is ineffective or not tolerated, intensification of therapy with additional medications (guided by consideration of the particular long QT syndrome type), left cardiac sympathetic denervation, and/or an ICD is recommended (2, 6-12).
I	B-NR	3. In patients with long QT syndrome and recurrent appropriate ICD shocks despite maximum tolerated doses of a beta blocker, intensification of medical therapy with additional medications (guided by consideration of according to the particular long QT syndrome type) or left cardiac sympathetic denervation, is recommended (6, 7, 10, 13-16).
I	B-NR	4. In patients with clinically diagnosed long QT syndrome, genetic counseling and genetic testing are recommended (17-21).
IIa	B-NR	5. In patients with suspected long QT syndrome, ambulatory electrocardiographic monitoring, recording the ECG lying and immediately on standing, and/or exercise treadmill testing can be useful for establishing a diagnosis and monitoring the response to therapy (22-29).
IIa	B-NR	6. In asymptomatic patients with long QT syndrome and a resting QTc less than 470 ms, chronic therapy with a beta blocker is reasonable (3, 30, 31).
IIb	B-NR	7. In asymptomatic patients with long QT syndrome and a resting QTc greater than 500 ms while receiving a beta blocker, intensification of therapy with medications (guided by consideration of the particular long QT syndrome type), left cardiac sympathetic denervation or an ICD may be considered (2, 8, 11, 30).
III: Harm	B-NR	8. In patients with long QT syndrome, QT-prolonging medications are potentially harmful (5, 12, 32-34).

2017 AHA/ACC/HRS guideline for management of patients with ventricular arrhythmias and the prevention of sudden cardiac death

Figure 9. Prevention of SCD in Patients With Long QT Syndrome



누나와 어머니를 포함 외삼촌과 이모의 대부분이 돌연사한 무증상의 20세 남성



2017 AHA/ACC/HRS guideline for management of patients with ventricular arrhythmias and the prevention of sudden cardiac death

Recommendations for Catecholaminergic Polymorphic Ventricular Tachycardia References that support the recommendations are summarized in Online Data Supplement 41.		
COR	**LOE**	**Recommendations**
I	B-NR	1. In patients with catecholaminergic polymorphic ventricular tachycardia, a beta blocker is recommended (1, 2).
I	B-NR	2. In patients with catecholaminergic polymorphic ventricular tachycardia and recurrent sustained VT or syncope, while receiving adequate or maximally tolerated beta blocker, treatment intensification with either combination medication therapy (e.g., beta blocker, flecainide), left cardiac sympathetic denervation, and/or an ICD is recommended (2-6).
IIa	B-NR	3. In patients with catecholaminergic polymorphic ventricular tachycardia and with clinical VT or exertional syncope, genetic counseling and genetic testing are reasonable (7).


Lifestyle modification
ABS
Sympathectomy
ß-Blocker
ICD