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# Treatment methods for Long QT syndrome

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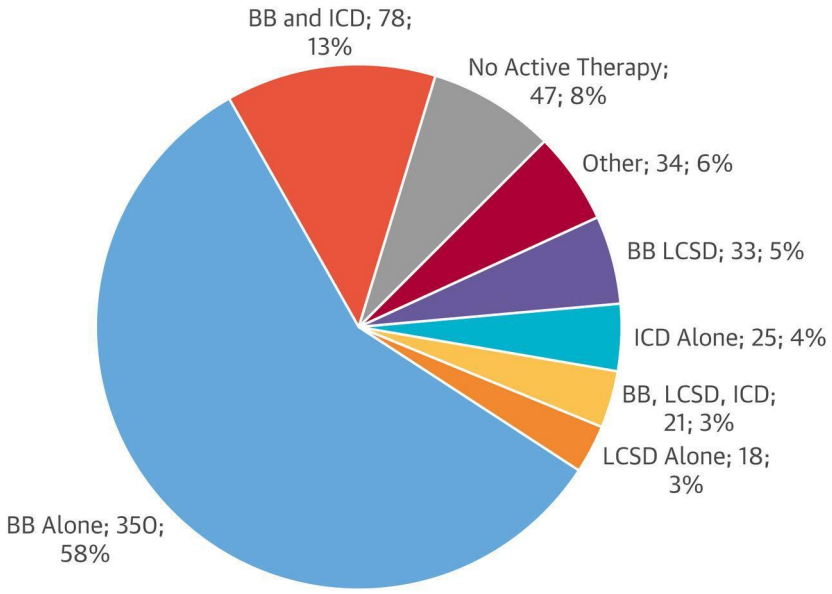
## Abstract
Long QT syndrome (LQTS) is a potentially fatal cardiac disorder caused by channelopathies. Such arrhythmia is often life threatening and might cause sudden cardiac death. There are many reasons of LQTS especially: specific medications and/or electrolytes imbalance or congenital causes like Jervell and Lange-Nielsen or Romano- Ward syndromes. In recent years many ways of treatment have been developed. Except conservative treatment with B-blockers also two surgical methods improved, implantable cardioverter defibrillator (ICD) and left cardiac sympathetic denervation (LCSD). Techniques of LCSD evolved causing side effects more tolerable and lowering the risk of Horner's syndrome.
Aim of this study is to review the role of sympathectomy in LQTS.

**Key words:** Sympathectomy, long QT syndrome (LQTS), B-blockers, Horner's syndrome.

## Introduction
Long QT syndrome (LQTS) is a group of cardiac arrhythmias characterized by delayed cardiac repolarization and improved risk of sudden cardiac death. [1]. QT prolongation may be caused by many following factors, hypokalemia, hypocalcemia or hypomagnesemia and QT-prolonging drugs namely antihistamines, antiarrhythmic, antibiotics, antidepressants. [2-4]. Diagnosis of LQTS requires 12 lead ECG and measurement QT interval. For the further examination the Bazett's formula is being used. Prolongation of QTc above 450ms (milliseconds) in men and 460ms in women should be regarded as abnormal [5]. Main reason of congenital LQTS are genes mutations of which 80% are being caused by three major mutations [6].

## Main body
There are three common LQTS treatment ways, such as B-blockers, implantable cardioverter defibrillator (ICD) and left cardiac sympathetic denervation (LCSD). Most common way of treatment is pharmaceutical with B-blockers. Patients with complex ventricular arrhythmias with family history of early sudden cardiac death or Qtc interval over 500ms B-blockers like propranolol or nadolol are recommended in maximally tolerated doses [7,8]. A study based on an electronic questionnaire sent out to the European Heart Rhythm Association (EHRA) showed that the first-line

therapy in LQTS is medication (76%) followed by a combination of drugs and implantable cardioverter defibrillator (ICD) (19%) [9]. Indications for ICD implantation are patients with diagnosed LQTS who survived cardiac arrest (Class I) or patients with LQTS who suffer recurrent arrhythmias despite high dose B-blocker therapy (Class IIa) [10].
The role of LSCD is proven to be effective but rarely performed. Among retrospective studies there were 85 cases of LSCD use [11]. Schwartz et al. Concluded that LSCD is an effective therapy for patients with recurrent arrhythmias despite B-blockers therapy. After 13 years of studies there were 147 patients with LCSD identified and treated. [12]. Final statement which they proposed was, that LCSD should be considered for patients with recurrent arrhythmia episodes under maximal pharmacological treatment and for patients who suffer arrhythmia despite the presence of an ICD.




**[Figure 1]. Treatement of LQTS summary.**

Recommendations of the EHRA for LCSD are patients with diagnosis of LQTS in whom: (I) ICD therapy is contraindicated or refused and/or (II) B-blockers are either not effective in preventing syncope/arrhythmias, not tolerated, not accepted or contraindicated (Class I) and it may be useful in patients with a diagnosis of LQTS who experience breakthrough events while on therapy with B-blockers/ICD (Class IIa)[10,11], [Figure 1].
Ways of performing left cardiac sympathetic denervation have evolved over the years. Stellectomy and cervicothoracic sympathectomy of left side is not being performed any longer due to a high amount of side effects such as Horner's syndrome and very little cardiac denervation. Nowadays the hight ablation of lower third thoracic ganglia together with ganglia T2 to T4 is being performed. Such technique provides high percentage of good outcomes and low percentage of side effects, rarely causing Horner's syndrome. [13]. Also there is the new mini invasive treatment, video-assisted thoracoscopic LCSD (VATS-LCSD) developing, causing no need for open thoracotomy. [14]. However there are following side effects of such treatment, dry left arm, face and forehead with intense sweating of the right side. Horner's syndrome or ptosis is now rare and mostly temporary [15].

## Conclusion

Both medical and invasive treatment of LQTS prove fair results. Common treatment with B-blockers is obvious due to a costs and availability of such treatment. However there are some recommendations for invasive treatment for patients in risk groups developing high risk factors. ICD and LCSD are methods being strongly improved among the years. Future studies may prove that invasive treatment might play a key role in both treatment and prevention of sudden cardiac death. LCSD has an significant role in such treatment maintaining high amount of good outcomes with minimal long term complications. Careful clinical selection of patients for LCSD is crucial.
It is said that in future improved LCSD might be both cheaper and more effective way of treatment of LQTS than ICD.

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