HRS/EHRA [2013] Expert Consensus Statement: **LQTS**

*Expert Consensus Recommendations on **LQTS Diagnosis***

1. LQTS is diagnosed: a. In the presence of an LQTS risk score ≥ 3.5 in the absence of a secondary cause for QT prolongation, *and/or* b. In the presence of an unequivocally pathogenic mutation in one of the LQTS genes, *or* c. In the presence of a QTc≥ 500 ms in repeated 12-lead ECG and in the absence of a secondary cause for QT prolongation.
2. LQTS can be diagnosed in the presence of a QTc between 480-499 ms in repeated 12-lead ECGs in a patient with unexplained syncope in the absence of a secondary cause for QT prolongation and in the absence of a pathogenic mutation.

Heart Rhythm (2013), in press (ePUB)
Europace (2013), in press

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Expert Consensus Recommendations on ***LQTS Therapeutic Interventions***

Class I	1. The following lifestyle changes **are recommended** in all patients with a diagnosis of LQTS: a) Avoidance of QT prolonging drugs (www.qtdrugs.org) b) Identification and correction of electrolyte abnormalities that may occur during diarrhea, vomiting, metabolic conditions or imbalanced diets for weight loss. 2. Beta-blockers **are recommended** for patients with a diagnosis of LQTS who are: *a)* Asymptomatic with QTc ≥ 470 ms, *and/or* b) Symptomatic for syncope or documented VT/VF. 3. Left cardiac sympathetic denervation (LCSD) **is recommended** for high-risk patients with a diagnosis of LQTS in whom: a) ICD therapy is contraindicated or refused, *and/or* b) Beta-blockers are either not effective in preventing syncope/ arrhythmias, not tolerated, not accepted or contraindicated. 4. ICD implantation **is recommended** for patients with a diagnosis of LQTS who are survivors of a cardiac arrest. 5. All LQTS patients who wish to engage in competitive sports **should be** referred to a clinical expert for evaluation of risk.

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Class IIa	6. Beta-blockers **can be useful** in patients with a diagnosis of LQTS who are asymptomatic with QTc ≤ 470ms.
	7. ICD implantation **can be useful** in patients with a diagnosis of LQTS who experience recurrent syncopal events while on beta-blocker therapy. 8. LCSD **can be useful** in patients with a diagnosis of LQTS who experience breakthrough events while on therapy with beta-blockers/ICD. 9. Sodium channel blockers **can be useful**, as add-on therapy, for LQT3 patients with a QTc > 500 ms who shorten their QTc by > 40 ms following an acute oral drug test with one of these compounds.
Class III	10. Except under special circumstances, ICD implantation **is <u>not</u> indicated** in asymptomatic LQTS patients who have not been tried on beta-blocker therapy.

Heart Rhythm (2013), in press (ePUB)
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- Mutation detection rate (test sensitivity): 75-80%

I Recommended	Strong clinical suspicion for LQTS Asymptomatic long-QT intervals (QTc: >500 ms adults, >480 ms prepuberty) Mutation-specific testing in family members
IIa Can be useful	
IIb Can be considered	Asymptomatic long-QT intervals (QTc: 480-500 ms adults, 460-480 ms prepuberty)
III Not recommended	
Gene **A** >10% **B** 1-10% **C** <1%	**LQT-1 (KCNQ1), LQT-2 (KCNH2)** **LQT-3 (SCN5A)** LQT-4 to LQT-13

HRS/EHRA [2011] Expert Consensus Statement: **Family Members**

- Mutation-specific testing is recommended for family members **(first degree blood relatives)** in nearly ALL familial heart diseases after identification of a mutation

I Recommended	Genetic test results in therapeutic recommendation (e.g., significant life-style modification or directed therapy)
IIa Can be useful	Diagnostic clarification (e.g., increased clinical surveillance, early interventions and prevention), Clarification or confirmation of disease etiology, Exclusion of genetic disease determination and development
IIb Can be considered	
III Not recommended	

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