Expert Consensus Recommendations on LQTS Therapeutic Interventions

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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 in patients with a diagnosis of LQTS who are: a. Asymptomatic with QTc ≥470 ms and/or b. Symptomatic for syncope or documented ventricular tachycardia/ventricular fibrillation (VT/VF).

3. Left cardiac sympathetic denervation (LCSD) is recommended in high-risk patients with a diagnosis of LQTS in whom: a. Implantable cardioverter-defibrillator (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 in 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 the evaluation of risk. Class IIa

6. Beta-blockers can be useful in patients with a diagnosis of LQTS who are asymptomatic with QTc ≤470 ms.

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 4500 ms who shorten their QTc by 440 ms following an acute oral drug test with one of these compounds.

10. Class III 10. Except under special circumstances, ICD implantation is not indicated in asymptomatic LQTS patients who have not been tried on beta-blocker therapy.

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 corrected QT interval for heart rate using Bazett's formula (QTc) ≥500 ms in repeated 12- lead electrocardiogram (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 and 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.

Priori SG, Wilde AA, Horie M, et al. ; Heart Rhythm Society; European Heart Rhythm Association; Asia Pacific Heart Rhythm Society.Executive summary: HRS/EHRA/APHRS expert consensus statement on the diagnosis and management of patients with inherited primary arrhythmia syndromes.Europace. 2013 Oct;15(10):1389-406.