

What is Long QT syndrome?

Long QT syndrome (LQTS) is a cardiac electrophysiologic disorder, characterized by QT prolongation and T-wave abnormalities on the ECG that are associated with tachyarrhythmias, typically the ventricular tachycardia *torsade de pointes*. It is a genetic condition, and the three most common types include:

- Type 1 (LQT1) – symptoms tend to be triggered by exercise
- Type 2 (LQT2) – symptoms tend to be triggered by stress or sudden or unexpected sounds
- Type 3 (LQT3) – symptoms tend to occur while resting or asleep

Symptoms include pre syncope, syncope, palpitations, and dizziness. Beta-blockers are the first line treatment for LQTS. Children commenced on beta-blockers are likely to need lifelong treatment.

Which beta-blockers are used to treat LQTS?

Choice of treatment will be identified by clinical specialist. Propranolol (licensed in children) and nadolol are the most effective for long QT. Metoprolol is less effective and should be avoided. **Propranolol** is first choice treatment in young children due to its shorter half-life and availability of a liquid preparation. Propranolol has a better QTc-shortening effect compared to metoprolol and nadolol¹. Please refer to the latest BNFC for dosing for arrhythmias.

Atenolol (licensed in >12 years) is preferred in school-aged children where once or twice daily dosing is beneficial - follow BNFC for dosing.

Nadolol and bisoprolol (both unlicensed in children) are preferred in older children/ adolescents, where once or twice daily dosing is convenient (increased half-life). Nadolol is associated with a significant risk reduction in patients with LQT2 comparative to other beta-blockers². Bisoprolol has less evidence but comparable so far; the same effectiveness but reduced risk of adverse effects³.

Nadolol dosing depends on the age of the child and extent of symptoms. The only licensed preparation in the UK is an 80mg tablet hence dosing is rounded to the nearest 20mg (quarter tablet) to ease administration. The usual range is from 20-80mg once daily titrated to response. A starting dose of 0.5-1mg/kg once daily, can be used in younger children.

Bisoprolol dosing depends on the age and extent of symptoms. The usual starting dose is 1.25 – 2.5mg once daily, titrated to response.

Adverse drug effects with beta-blockers

If any adverse effects are reported, please contact the specialist team to discuss the next steps.

- Fatigue/tiredness/Cold peripheries – common when initiating and during dose increments. Expect to subside over one or two weeks. Contact the specialist if persists for more than two weeks
- Hypotension/dizziness – consider giving dose before bedtime, if problematic and encourage fluid and salt intake. If persistent for >2 weeks, a dose reduction may be needed – contact the specialist for advice
- Syncope – contact the specialist immediately
- Nightmares – if severe, contact the specialist within a week for advice to consider switching to another beta blocker (e.g., water-soluble beta blocker e.g., nadolol)
- Shortness of breath – Consider underlying asthma diagnosis and please refer to the specialist, a cardioselective beta-blocker may be more appropriate.
- If a child develops asthma - please refer to the specialist, a cardio selective beta-blocker may be more appropriate. Do not stop the beta blocker.

When GPs will be asked to prescribe treatment?

GPs will be requested to take over treatment once a stable dose is established after the first follow up as a minimum which will be communicated to them via a clinic letter. Any dose changes or other treatment alterations will be communicated to GPs. Please notify parents if there is a change in concentration of preparation.

Monitoring treatment

All monitoring (such as Holters and ECG) to be performed during hospital appointments. GPs are not required to check ECG for appropriateness or safety.

Contact details

Royal Brompton ICC specialist team: 0330 128 8121 ext 82205 (Paediatrics)

Email: gstt.hlcc.iccnurses@nhs.net

Evelina cardiology specialist team: 020 7188 4000 (ext 58389)

Email: gstt.paediatriccardiologycns@nhs.net

GSTT Pharmacy MI helpline: 020 7188 88750

Email: gstt.mymedicines@nhs.net

References

1. Chockalingam P, Crotti L, Girardengo G, Johnson J, Harris K, van der Heijden J et al. Not All Beta-Blockers Are Equal in the Management of Long QT Syndrome Types 1 and 2. *Journal of the American College of Cardiology*. 2012;60(20):2092-2099.
2. Abu-Zeitone A, Peterson D, Polonsky B, McNitt S, Moss A. Efficacy of Different Beta-Blockers in the Treatment of Long QT Syndrome. *Journal of the American College of Cardiology*. 2014;64(13):1352-1358.
3. Fazio G, Vernuccio F, Lo Re G, Grutta G, Mongioli M. Role of Bisoprolol in Patients with Long QT Syndrome. *Annals of Noninvasive Electrocardiology*. 2013;18(5):467-470.