Reference Number: UHS0BS191 **Version Number: 3**	**Date of Next Review: 18/6/27** **Previous Trust/LHB Reference Number:**

Long QT Syndrome

Introduction and Aim

Long QT Syndrome is an abnormality of cardiac repolarisation which may be inherited or acquired.
Predisposes to the development of malignant ventricular arrhythmias triggering syncope, seizures and sudden cardiac death in an otherwise healthy individual
In pregnancy there is an increased chance of arrhythmia with the **highest risk postpartum**.

Scope

Equality Health Impact Assessment	*An Equality Health Impact Assessment (EHIA) has not been completed.*
Approved by	*Maternity Professional Forum and Obstetrics & Gynaecology Quality & Safety* Cardiff and Vale University Health Board Safeguarding Steering Group Meeting
Accountable Executive or Clinical Board Director	*Jason Roberts, Executive Nurse Director*
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Summary of reviews/amendments

Version Number	Date of Review Approved	Date Published	Summary of Amendments
1	2016		
2	2020		
3	2024		

"We recognise maternity and gynaecological services will be accessed by women, gender diverse individuals and people whose gender identity does not align with the sex they were assigned at birth. Therefore, we believe delivery of care must at all times be appropriate, inclusive and sensitive to the needs of everyone." (RCOG, 2022)

Aim to continue beta blockers throughout pregnancy and particularly in the postpartum period.

Incidence	1 in approximately 8 000
Presentation:	Family history, or may be index case Arrhythmia- palpitations, atypical syncope especially during exercise Characteristic ECG abnormality with cardiac symptoms
Precipitated by:	Certain drugs, electrolyte abnormalities, hypothermia, exercise, emotion
Diagnosis:	Genetic testing if a carrier of a recognised mutation (30% of people with long QT do not have a recognised mutation) ECG – (resting ECG may be normal). Duration of Q-T interval corrected for heart rate (QTc) is best prognostic indictor Longer QTc times are associated with higher risk.
ECG:	**Prolonged QTc >470 ms in adult females** (Bazzett's formula)
Anatomy:	Structurally normal heart
Treatment:	**Main stay of treatment is beta blocker therapy** which reduces the chances of sudden cardiac death. High risk individuals may be fitted with an implantable cardiac defibrillator (ICD)

Inherited

Genetic abnormality affecting specific of K+ Na+ or Ca2+ cardiac channels all affecting cardiac repolarisation Most types autosomal dominant

12 different genotypes described; types 1,2 and 3 being the commonest.

Type	Genotype	Channelopathy	Incidence	Highest Risk
1	KCNQ1	K+	45%	Adrenergic stress, exercise
2	KCNH2	K+	40%	Adrenergic stress, increased risk post-partum
3	SCN5A	Na2+	5%	More common during rest/sleep
4 - 12			rare	

Acquired

Often manifests in those with underlying genetic predisposition
iatrogenic- resulting from effect of
QT prolonging medication
Metabolic e.g. severe anorexia nervosa

- Consider referral to Clinical Genetics for counselling and genetic testing
- Beta blockers (e.g. nadolol) are extremely protective in LQT1 patients and moderately protective in LQT2 and LQT3. Awareness of LQTS genotype allows selective use of Beta blockers. Beta blocker medication throughout pregnancy is associated with fetal growth restriction so offer fetal surveillance by serial growth scans (alternatively consider post- partum treatment).
- High risk individuals may be fitted with an implantable cardiac defibrillator (ICD) - arrange an ICD check & download
- **Avoid QT prolonging medication—please refer to full list at back of high risk folder at reception on delivery suite or see www.crediblemeds.org**
- Risk of congenital LQTS with fetal heart block. Monitor fetal heart rate by auscultation or ultrasound every 4 weeks from 20 weeks gestation. Refer for fetal echo if persistent fetal bradycardia <110bpm.

Peri-Partum Care

Continue beta blockers peripartum

Perform baseline ECG on admission and repeat if concerning symptoms of palpitations/ syncope

Check U&E's- Maintain K+ >4.5, correct hypomagnesaemia and hypocalcaemia

Minimise sympathetic stress, consider early effective epidural & good postop pain control

Ensure defibrillator is readily available if ICD not in situ

If ICD is fitted do not inactivate for labour. Avoid unipolar diathermy in theatre.

If unipolar diathermy must be used inactivate the ICD by securing a clinical magnet over the device (kept in cardiac arrest trolley on DS, theatre end). Fit defib pads while ICD inactivated.

Avoid hypothermia- measure temp and institute active warming if indicated

Avoid QT prolonging medication- for up to date information see www.crediblemeds.org

Drugs specific to pregnancy

This list is not exhaustive. Please check other medications at ***www.crediblemeds.org***

Antiemetics	Dexamethasone, cyclizine safe. **Avoid** Ondansetron, metoclopramide
Analgesia	Morphine, paracetamol, NSAIDs – safe. Tramadol—avoid as possible risk
Antibiotics	Augmentin and cefuroxime safe. **Check other antibiotics.**
Tocolytics	Avoid terbutaline, GTN is safe alternative
Regional anaesthesia	Safe
Vasopressors	Phenylephrine/metaraminol first choice, avoid ephedrine if possible
GA induction	Opiates safe- give short acting to obtund the pressor response to intubation. Thiopentone- safe, Propofol- probably safe Rocuronium and sugammadex- safe. **Avoid** Suxamethonium and Neostigmine/glycopyrrolate Sevoflurane controversial- change to isoflurane if time allows, **if not proceed**
GA maintenance	Avoid hypercapnoea. N2O- safe

Malignant Arrythmias

- **Manage according to advanced life support guidelines and defibrillate if indicated**
- Use Magnesium sulphate to prevent recurrence of arrhythmia. Use 2g over 3-4 minutes
- Loading dose, may be repeated after 15 minutes. Loading dose may be used prophylactically in high risk situations.
- Supplement K+ to >4.5mM/L
- Consider trans-venous pacing to increase heart rate in recurrent Torsades
- Avoid QT prolonging antiarrhythmics- amiodarone and flecainide contraindicated

References

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- Crediblemeds.org.uk accessed 17/11/20

- C. Staikou, K. Chondrogiannis and A. Mani (2012) Perioperative management of hereditary arrhythmogenic syndromes British Journal of Anaesthesia. 108 (5): 730-744. doi: 10.1093/bja/aes105