

Recommendations for the management of patients with Long QT syndrome in dentistry

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Abstract

Aim and objectives: Long QT syndrome (LQTS) is a rare but potentially life threatening disorder of the heart's conduction system, where cardiac repolarisation (reflected in the QT interval on the ECG) is prolonged. It may be congenital or acquired. Patients may experience palpitations, syncope, or seizure-like episodes due to *torsades de pointes* (ventricular tachycardia). Arrhythmias can lead to cardiac arrest or sudden cardiac death. Physical activity, auditory stimuli, emotional stress or drugs can provoke arrhythmias. We aim to improve awareness of LQTS and have developed recommendations using available evidence and supporting professional opinion.

Methodology: A review of the literature was carried out as the first stage of guideline development, and a multidisciplinary guideline development group was established to develop the recommendations.

Results: We have developed recommendations endorsed by the UK Association for Inherited Cardiac Conditions (AICC) recommending that these patients receive their dental care in general dental practice with adherence to practical key advice and recommendations. Dentists need improved awareness of the disease, and the list of drugs which are contraindicated to avoid arrhythmic complications. Patients may be referred to secondary or tertiary dental services based on their dental needs in the same way as patients without LQTS would.

Conclusions: Detailed guidance on the dental management of patients with LQTS is lacking, but, using available evidence with supporting professional opinion, we have developed recommendations for the dental management of patients with LQTS in the UK. This paper outlines the rationale and processes involved in the development of this guidance.

Key words: Long QT syndrome, LQTS, dentistry

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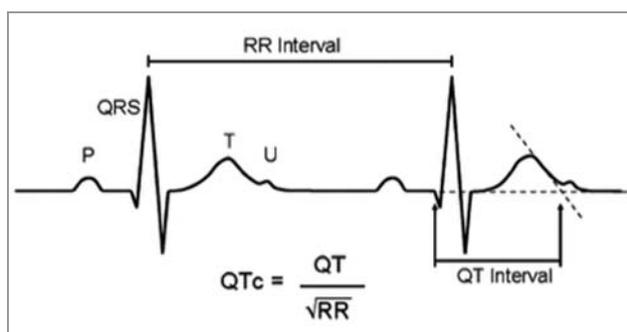
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Introduction

Long QT syndrome (LQTS) is a disorder of the conduction system characterised by heterogeneity of cellular repolarisation, which can result in ventricular tachyarrhythmia. The diagnosis is based on assessment of the length and morphology of the QT interval on the

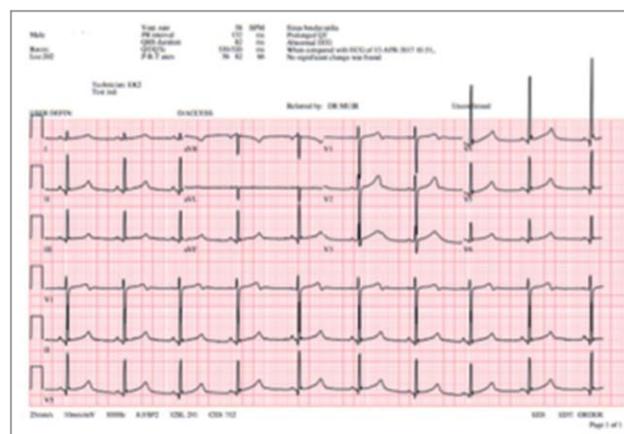
electrocardiogram (Schwartz and Crotti, 2011) (Figure 1), but secondary causes of an acquired QT-prolongation, such as exposure to drugs, metabolic abnormalities or electrolyte imbalances should be excluded. Congenital LQTS is a rare condition with an estimated incidence in the Caucasian population of 1:2000 live births (Schwartz et al., 2009).

Figure 1: Measuring the QT interval (adapted from Johnson and Ackerman, 2009)



Traditionally, patients with LQTS in Northern Ireland have been treated in general dental practice or referred to the hospital dental services for monitoring during dental treatment, but the level of monitoring that patients receive is widely variable. Knowledge of the inherited disorders of conduction, such as LQTS, is limited among dental practitioners across general dental practice, community and hospital settings. One Welsh study of 528 dentists reported that up to 76% of all practitioners were not aware of the appropriate management of LQTS for dental procedures (Thompson *et al.*, 2007). The evidence-base in the literature for the dental management of patients with LQTS is limited, although key points and recommendations can be identified. Dentists need to avoid certain drugs, including adrenaline, to avoid arrhythmic complications. Improved awareness of the condition and its management will allow safe delivery of

Figure 2: LQTS ECG



dental care, reduce adverse events, and avoid unnecessary referral for monitoring during treatment. Clear guidance for the management of these patients does not exist locally or nationally, and through multidisciplinary team meetings and a review of the literature, regional guidance has been developed for the management of patients with LQTS.

What is LQTS?

The congenital LQTS is a rare, inherited cardiovascular condition characterised by abnormal cardiac repolarisation due to malfunctioning cardiac ion channels. The clinical

Figure 3: Schwartz Score

Characteristics	Score
Electrocardiographic findings	
• QTc (calculated with Bazett formula)	
– ≥480 ms	3
– 460-470 ms	4
– 450 ms and male gender	1
• QTc fourth minute recovery from exercise stress test ≥ 480 ms	1
• Torsades de pointes	2
• T wave alternans	1
• Notch T wave in three leads	1
• Low heart rate for age (children), heart rate below 2nd percentile for age	0.5
Clinical history	
• Syncope	
– With stress	2
– Without stress	1
• Congenital deafness	0.5
Family history	
• Other family members with definite LQTS	1
• Sudden death in immediate family members (before age 30 years)	0.5

SCORE ≤ 1 point – low probability LQTS; 1.5-3 points – intermediate probability LQTS; ≥ 3.5 points – high probability LQTS

manifestation of the condition can include arrhythmic events, due to *torsades de pointes* (ventricular tachycardia), which can result in syncope, or if prolonged, deterioration into ventricular fibrillation, cardiac arrest and/or sudden death. *Figure 2 (previous page)* illustrates the ECG changes seen in LQTS.

In 2015, the European Society of Cardiology published guidelines for the management of patients with ventricular arrhythmias and the prevention of sudden cardiac death, which included expert consensus recommendations for the diagnosis and management of patients with LQTS (Priori *et al.*, 2015). The diagnosis of the congenital LQTS is based primarily on the measurement of the corrected QT interval (QTc) on the surface ECG. A scoring system, known as the Schwartz score (*Figure 3 - previous page*), takes into account other ECG features as well as the personal and family history of the individual and provides a probability of the diagnosis of congenital LQTS (Schwartz and Crotti, 2011).

Secondary causes of QTc prolongation must be excluded prior to a diagnosis of congenital LQTS. These include certain drugs, electrolyte or metabolic abnormalities and other acquired cardiac conditions. Table 1 lists drugs encountered in dental practice known to cause QTc prolongation. For up-to-date information on QTc prolonging drugs it is recommended to consult a web based resource such as the British National Formulary UK (www.bnf.nice.org.uk), CredibleMeds® (www.CredibleMeds.org) (Woosley *et al.*, 2013) or the registered charity organisation Sudden Arrhythmic Death Syndrome (SADS) (www.sads.org.uk) (Cox, 2018).

Medical management

The medical management of LQTS aims to control symptoms and prevent sudden cardiac death. Lifestyle modifications are recommended routinely to all LQTS

patients. These include the avoidance of strenuous sporting activity and potentially QT prolonging drugs (*Table 1*). Specific therapies for LQTS include beta-blockers, implantable cardioverter defibrillators (ICDs) and left cardiac sympathetic denervation (LCSD). Beta-blockers are currently recommended in asymptomatic patients with LQTS with a QTc ≥ 470 msec and/or symptomatic patients with a history of syncope or documented ventricular arrhythmia. They can be useful in patients with a diagnosis of LQTS who are asymptomatic and have normal QT intervals of ≤ 470 msec (Priori *et al.*, 2015) and are reported to prevent cardiac events in approximately 70% of patients (Kahn, 2002).

ICD implantation is recommended for all patients with LQTS who survive a cardiac arrest and prophylactic implantation is recommended in high-risk individuals, including those with breakthrough events on medical therapy. LCSD is recommended in high-risk patients with LQTS in whom ICDs are contraindicated or refused, and/or patients in whom beta-blockers have failed to control cardiac tachyarrhythmias (Priori *et al.*, 2015).

Material and method

Review of current practice

A review of the management of this group of patients in Northern Ireland revealed that most received their treatment in general dental practice, while a smaller number were referred to hospital dental services for treatment with clinical monitoring. The level of monitoring provided in the hospital ranged from visual monitoring of the patient during the procedure to monitoring by an anaesthetist in a theatre environment with a 3-lead ECG. The need for standardised guidance for the dental management of this group of patients was identified, and a series of multidisciplinary team meetings followed.

Table 1: QTc prolonging drugs

Drug Group	Drug Name
Catecholamine	Adrenaline/epinephrine
Antidepressants	Amitriptyline, Citalopram, Fluoxetine, Sertraline
Antibiotics	Azithromycin, Erythromycin, Metronidazole
Anti-emetics	Domperidone, Ondansetron
Antifungals	Fluconazole, Itraconazole, Ketoconazole
Antihistamines	Diphenhydramine
Antihypertensives	Furosemide, Isradipine, Indapamide, Nifedipine
Antiarrhythmics	Amiodarone, Disopyramide, Quinidine, Sotalol
Psychiatric drugs	Chlorpromazine, Clozapine, Droperidol, Phenothiazide, Haloperidol, Lithium, Methadone, Risperidone, Venlafaxine
Asthma drugs	Albuterol, Formoterol, Salbutamol, Salmeterol, Terbutaline
Chemotherapy drugs	Tamoxifen
HIV/AIDS drugs	Nelonavir, Rilpivirine, Fosarnet, Ritonavir, Saquinavir

NB. This list is not exhaustive, and highlights only some commonly encountered drugs in general practice. An up-to-date web based list such as the BNF www.bnf.nice.org.uk, www.CredibleMeds.org, or www.sads.org.uk should be consulted before prescribing.

A review of the literature was carried out as the first stage of guideline development using multiple search phrases for LQTS and dentistry in MEDLINE and EMBASE. Only four papers relating to the dental management of patients with LQTS were identified (Ahmed and Elseed, 2005; Karp and Ganoza, 2006; Karp and Moss, 2006; Rochford and Seldin, 2009).

Recommendations identified from the literature are listed below:

- Beta-blocker therapy should be continued as normal on the day of treatment (Brooker *et al.*, 2003; Staikou *et al.*, 2012)
- Alleviate patient anxiety. Pre-medication with oral benzodiazepines can be administered where required. Nitrous oxide and midazolam are safe for this group (Michaloudas *et al.*, 1995; Michaloudas *et al.*, 1996). Topical anaesthetic should be used appropriately to minimise pain and anxiety prior to the administration of an appropriate local anaesthetic. A quiet, calm environment is recommended, but in the reality of the dental surgery, it is recommended to keep noise to a minimum with phones turned off and avoidance of sudden loud noises, where possible. Music therapy, hypnosis and cognitive behavioural therapy may also be considered in the treatment of dental anxiety or phobia
- Avoid drugs known to prolong the QT interval (see *Table 1*). As adrenaline can prolong the QT interval, its use in local anaesthetic is contraindicated (Sun *et al.*, 1998; Ackerman *et al.*, 2002; Vyas *et al.*, 2006; Karp and Ganoza, 2006; Karp and Moss, 2006; Rochford and Seldin, 2009) but alternative vasoconstrictors can be used. Bupivacaine can induce arrhythmias and is best avoided (Cucchiari and Rhodes, 2003)
- Electrolytes should be checked and corrected if necessary before general anaesthetic (GA) to ensure normal cardiac repolarisation (Brooker *et al.*, 2003). It is important to assess serum electrolytes prior to surgery. In particular, potassium, magnesium and calcium deficiencies should be identified and replaced where necessary prior to surgical dental treatment (Brooker *et al.*, 2003)
- Where patients have a pacemaker or ICD fitted, it is important to avoid electrosurgery.

The concern with some dental equipment and ICDs relates to electromagnetic interference (EMI), which can potentially cause malfunctions in the device. A number of investigators have examined the effects of a variety of dental instruments both *in vitro* and *in vivo*. Electrosurgery can sometimes be used in dentistry to remove soft tissue in cases of gingival overgrowth or to provide crown lengthening. The instruments commonly used are monopolar and so they are earthed by way of a metal plate often placed behind the patient's back. The complication of a current interfering with an ICD is therefore high. Most authors agree that the use of electrosurgery above the level of the umbilicus has therefore the potential to cause significant interference with ICDs and pacemakers (Crossley *et al.*, 2011). Although the use of electrosurgery in general dental practice is relatively infrequent, it would appear that the continued advice should be to avoid its use in patients with ICDs and pacemakers.

Maiorana *et al.* (2013) concluded that piezo electric ultrasonic dental scalers had no ill effect on ICDs in an *in vivo* experiment involving 12 patients. Other studies with *in vitro* experimental designs did report some interference particularly when equipment was in very close proximity to ICDs, although this did not cause any significant functional complications (Maheshwari *et al.*, 2015). There is evidence to show that commonly used dental equipment such as curing lights, apex locators, endodontic heat guns and dental scalers are safe to use with these patients (Garofalo *et al.*, 2002; Trenter and Walmsley, 2003; Lahor-Soler *et al.*, 2015).

In addition to the above literature review on the use of dental equipment interference with ICDs, a local Consultant Cardiologist also investigated this further with the Regional Cardiac Devices Laboratory in Northern Ireland, who confirmed that these devices may be used safely, adding that the manufacturers' instructions should always be checked before use. Further research in this area would provide added clarification. It is essential to have cardio-pulmonary resuscitation (CPR) and basic life support (BLS) skills up to date in the event of ventricular tachyarrhythmia and cardiac arrest. Access to an automated external defibrillator (AED) within a reasonable time frame is essential.

In Northern Ireland the RQIA (Regulation and Quality Improvement Authority), equivalent to the Care Quality Commission (CQC) in England, state that dental practitioners have access to an AED within a reasonable time frame (RQIA, 2005). When contacted to clarify this they stated dental practitioners should use their own judgement and risk assessment to determine whether they need to have an AED in their practice. The CQC website (CQC, 2018) states that they expect a practice to follow the national guidance issued by the Resuscitation Council, and where an AED is not available immediately, they would expect to see a robust and realistic risk assessment detailing how an AED could be accessed in a timely manner, as the emergency services may not always be able to respond in the critical first few minutes of an acute cardiac arrest. The GDC Standards for the Dental Team state that, as a dental professional, you must follow the guidance on medical emergencies and training updates issued by the Resuscitation Council (GDC, 2017). The Resuscitation Council (2018) expects that all clinical dental areas should have immediate access (within the first minutes of a cardiorespiratory arrest) to oxygen, resuscitation equipment for airway management, and an AED. Therefore, we recommend that dental practices carry out an individual risk assessment in line with their regulatory body requirements to determine if they need an AED on the dental premises.

A careful review of the medical history by the dental practitioner should elicit a known history of LQTS prior to dental therapy, indicating that precautions are necessary.

As an initial informal survey to gauge how LQTS was managed elsewhere in the UK and Ireland, the authors contacted a random sample of 14 UK based Special Care Dentistry Consultants as a sample of practitioners who may most often receive referrals of LQTS patients for dental management. Seven responses were received and revealed a wide range in treatment options: from normal outpatient treatment or IV sedation (even for the non-anxious), to admission for treatment under general anaesthetic. All

respondents agreed that ECG monitoring would require medical input for lead placement and interpretation. The Belfast Inherited Cardiac Conditions team also informally discussed the issue with colleagues across the UK to gauge consensus opinion and there was no clear consensus on management other than continue beta-blocker therapy and avoidance of contraindicated medications.

Whilst there is a spectrum of risk of arrhythmias in patients with LQTS, there is no evidence to prove that this reflects an individual's risk during dental treatment as all patients with LQTS have the potential for significant arrhythmias. This review of practice revealed that across the UK some patients with LQTS are being referred to secondary or tertiary dental services, whilst the majority are being managed in the primary dental service, despite limited awareness on how to manage the condition (Thompson *et al.*, 2007). The authors aimed to standardise care for patients with LQTS and provide dentists working in all areas of dentistry with the advice and recommendations required to avoid adverse events.

Results

The group concluded that it would be appropriate for LQTS patients to have their routine dental care in the primary or general dental services. The rationale was that most of these patients currently receive dental care in the general dental setting and, bar one patient known to cardiology to have experienced episodes of syncope at their dentist (in the setting of administration of QT prolonging medication), no adverse outcomes had been reported to the regional cardiology services. Referral to hospital based specialist dental services with unfamiliar surroundings in itself may induce anxiety. Dentists, and dental care professionals, undergo annual CPR/BLS training, training in medical emergencies, and dental practices should have access to an AED within a reasonable time frame as outlined previously. Patients with ICDs are not considered 'high risk', as the ICD offers best protection. As outlined previously, modern electronic dental equipment are unlikely to present these patients with any difficulties with the exception of monopolar electrosurgery units.

Discussion

Prevention of dental disease should be of paramount importance to reduce the need for intervention, with prompt treatment of dental pain and infection to minimise the need for administration of drugs or antibiotics which may prolong the QT interval. In a medical emergency, all necessary rescue medications should be given as usual. In the case of anaphylaxis, the risk of airway compromise poses a more immediate risk to the patient than that of potential ventricular tachycardia with administration of adrenaline, and management of anaphylaxis should be carried out as normal, with the history of LQTS highlighted when phoning for ambulance assistance.

In the rare scenario of a patient with LQTS requiring a procedure where there is the possibility of inability to gain

adequate anaesthesia without the use of adrenaline containing anaesthetic e.g. complex oral surgery or maxillo-facial procedures, referral to tertiary level specialist services should be made. There, the patient may undergo elective in-patient treatment with anaesthetic level monitoring. In select cases where anaesthesia may be predetermined to be complex, informed consent for the potential use of adrenaline containing medications can be sought. This requires careful planning and detailed discussion with the patient regarding the potential for adverse cardiac events.

These recommendations were presented, discussed and approved at both the Association for Inherited Cardiac Conditions (AICC) Annual General Meeting and Conference, and the Wales Gene Park, Cardiff International Cardiovascular Conference in 2015. Presentation of the recommendations as a poster at a national Special Care Dentistry conference, local dental audit, clinical effectiveness and continuing professional development meetings, and anaesthetic clinical effectiveness meetings highlighted no concerns with the recommendations, and guidance in this area was welcomed.

If the approach as presented is to be successful, it will involve targeted education of patients and dentists. Recommendations for the dental management of these patients are summarised in *Table 2*, and apply to both adults and children with a diagnosis of LQTS. This information will be disseminated to dentists in Northern Ireland and Continuing Professional Development lectures have been arranged.

For patients, education will be in the form of an information leaflet to include 3 key points (*Appendix 1*):

1. Take their beta blocker as normal on the day of their appointment;
2. Avoid adrenaline in local anaesthetic, and avoidance of contraindicated drugs;
3. Tell their dentist if they have an ICD.

Conclusion

LQTS is a rare but potentially life threatening condition. Detailed guidance on the dental management of patients with LQTS is lacking, but, using available evidence with supporting professional opinion, the authors have developed recommendations that have been endorsed by the Association for Inherited Cardiac Conditions (AICC) as recommended national guidelines for the dental management of patients with LQTS in the UK.

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Table 1: Recommendations for Dental Treatment

1	Adrenaline/epinephrine	
2	Avoid drugs known to prolong QT interval	<ul style="list-style-type: none"> – Contraindicated drugs in dentistry include: Antibiotics: Erythromycin, azithromycin, clarithromycin, metronidazole Antifungals: Fluconazole, itraconazole, ketoconazole
	Check up-to-date web based lists	<ul style="list-style-type: none"> – www.bnf.nice.org.uk – www.sads.org/living-with-sads/Drugs-to-Avoid – www.crediblemeds.org
3	Adrenaline is contraindicated	
4	Bupivacaine can induce arrhythmias and is best avoided	
5	Alleviate patient anxiety	<ul style="list-style-type: none"> – Effective analgesia; use topical anaesthetic – Consider conscious sedation <ul style="list-style-type: none"> • Nitrous oxide safe • Midazolam safe • Consider pre-medication with temazepam / diazepam – Quiet, calm environment
6	Check electrolytes prior to general anaesthetic	
7	Patients with ICD	<ul style="list-style-type: none"> – Electrosurgery can result in circuit damage, arrhythmia or burning – Modern ultrasonic scalers, apex locators and pulp testers may be used safely
8	Management of medical emergencies	<ul style="list-style-type: none"> – CPR / BLS training essential – Manage medical emergencies in the usual manner, including the administration of adrenaline in the case of anaphylaxis – Access to an automated external defibrillator (AED) as torsades (ventricular tachycardia) can deteriorate to VF and cardiac arrest NB: dental practices should carry out a risk assessment in line with their regulatory body requirements to determine if an AED is required on the premises.

NB. This list is not exhaustive, and highlights only some commonly encountered drugs in general practice. An up-to-date web based list such as the BNF www.bnf.nice.org.uk, www.CredibleMeds.org, or www.sads.org.uk should be consulted before prescribing.

Appendix 1: Patient advice leaflet

Recommendations for Dental Treatment in Patients with LQTS



Long QT Syndrome is a rare, inherited disorder of the heart's conduction system.

The diagnosis is made when QT interval (a measurement on the ECG) is prolonged.

This can put individuals at increased risk of heart rhythm disturbance which can lead to palpitations, black outs or rarely sudden death.

Management of the condition focuses on:

- Lifestyle modifications eg. avoidance of strenuous sporting activity and medications which can prolong the QT interval
- Beta blocker therapy to regulate the heart rhythm
- Internal Cardioverter-Defibrillators (ICD) in certain high risk groups

At the time of dental treatment individuals are at potential increased risk of abnormalities so simple precautions are necessary to reduce this.

Patients with LQTS should still receive their dental care in their general dental practice but the following guidelines should be followed:

1	Inform your dentist you have Long QT syndrome
2	Take your Beta Blocker as normal on the day of your dental treatment
3	Discuss with your dentist that certain medications should be avoided because of your LQTS (includes adrenaline in local anaesthetic and certain medications for infection)
4	Inform your dentist if you have an ICD

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