QTc prolongation in itself does not cause a ventricular arrhythmia. Instead, it is increased heterogeneity of repolarization that increases the risk of a polymorphic ventricular tachycardia such as Torsades de Pointes (TdP).

Diagnosis of congenital LQTS:
Characteristic ECG changes include prolonged QTc, torsades de pointes, T-wave alternans or notched T wave in at least three leads & a prolonged T_{p-e}. A history of syncope, congenital deafness, or a positive family history requires further investigation.

**LQTS genes:**
LQT1 patients have mutations on the KCNQ1 gene, which encodes the I_{Ks} current. LQT2 patients have mutations on the KCNH2 gene, which encodes the I_{Kr} current. LQT3 patients have mutations on SCN5A, the cardiac Na channel gene. LQT5 patients have mutations on the β-subunits of KCNQ1 (rare) & LQT6 patients have mutations on the β-subunits of KCNH2 (rare).

**Congenital & acquired LQTS:**
Accentuation of spatial dispersion of repolarization within the ventricular myocardium is the main arrhythmogenic substrate for both congenital & acquired LQTS. Spatial dispersion of repolarization is further exaggerated by sympathetic stimulation (especially in patients with LQT1 & 2). This exaggerated intrinsic heterogeneity, together with early after-depolarizations (EADs) or other aberrant electrical activity, may trigger the development of TdP. Risk factors for EADs include LQTS, hypokalaemia & sympathetic stimulation. Risk factors for drug-induced TdP include concurrent exposure to >1 drug with I_{Kr} blocking effect, cardiac disease, female gender, electrolyte abnormalities, bradyarrhythmias & genetic predisposition.
Anaesthetic drugs that prolong the QTc include all the halogenated inhalational agents, thiopentone, ketamine, glycopyrrolate, neostigmine, ondansetron, pancuronium & suxamethonium. However, none of these anaesthetic drugs increase repolarization heterogeneity. In contrast, catecholamines such as isoprenaline shorten the QTc of epicardial & endocardial cells but not M cells, so increase repolarization heterogeneity.

Anaesthetic management of child with known or suspected LQTS:
Liaise with the paediatric cardiologist & confirm the (likely) type of LQTS. If the patient is symptomatic, then temporary pacing should be considered. If a pacemaker or implantable defibrillator is already in situ, then check or revise the settings. Start β-blocking drugs if the child is not already taking them. Check that serum electrolyte concentrations (including magnesium) are normal; correct as necessary. Give oral midazolam 0.5 mg/kg as premedication. Monitor the ECG (2 leads) before, during & after surgery. Propofol or thiopentone should be used for induction of anaesthesia. Sympathetic stimulation during laryngoscopy & intubation must be minimised using a topical lidocaine spray or a bolus dose of alfentanil. Use a regional technique where appropriate to ensure good analgesia. Use cisatracurium if a relaxant is required; do not reverse, so use a nerve stimulator to monitor neuromuscular function. Avoid all risk factors for TdP. Have magnesium sulphate drawn up & a defibrillator in theatre.

Conclusions:
30% patients with LQTS have a normal ECG, so this problem is not as rare as you may think!
Susceptible patients may develop TdP after exposure to I_Kr blocking drugs, especially in the presence of halogenated anaesthetic agents. Magnesium sulphate is the drug of choice if a patient develops TdP. LQTS patients can be safely anaesthetised by avoidance of I_Kr blocking drugs, hypothermia & exogenous/endogenous catecholamines, & maintenance of normal serum electrolytes.

Key references: