With LQTS genetic testing, treatment strategies can be guided for men with LQT1 who have been asymptomatic (without symptoms) all their life (or for many, many years) may not need any active treatment, as their risk of developing symptoms at later ages is very low. As with all patients with LQTS, these low-risk patients need to avoid low blood potassium (caused by diuretic drug use, vomiting or diarrhea) and avoid (whenever possible) those drugs which aggravate the heart’s recharging system and prolong the QT interval. This combination of an otherwise dormant LQTS genetic substrate plus a ‘second hit’ can provide a fatal 1-2 punch. For a complete list of drugs that may be frequent. It is important to be sure that your child’s school is aware of this. And to consider obtaining a personal AED as part of the family’s LQTS safety gear. However, when well treated, remember that the AED should almost never be required.

It is important to be sure that your child’s school is aware of his or her condition and is properly prepared to respond in the event of an emergency. Visit StopSADS.org for care plans and other important materials that will help you in meeting with school personnel to ensure a safe environment for your child.

At the appropriate time, your child should also be informed that they can and should become a mother or father someday if they want to. Any recommendations that they should not become a parent because of their LQTS should be viewed as inappropriate nonsense. Pregnancy and delivery can be navigated successfully and relatively simply in women with LQTS but will require a teamwork approach between your LQTS specialist and the obstetrician.

Medication compliance

It is very important that LQTS-directed medical therapies be taken every day and not missed or omitted. The medications are not curative; they only provide protection while being taken and depending on the beta blocker, the protective effect can disappear within a day. After that the risk of cardiac events is the same as if the patient had not taken the medication at all. In addition, dose skipping/missing can actually be dangerous as the heart can become hypersensitive when the medication is not present.

Parents should teach their children about the importance of daily medication, and should make sure each daily dose is taken. Physicians need to discuss this directly with all patients, but particularly pre-teens and teenagers. The most common reason for cardiac events while on medication is that the medication was missed or stopped.

How can parents protect their kids?

Make sure the children take their medication daily, no missing doses.

See the doctor regularly for follow-up. Growing children need medication dose changes regularly. Make sure you see the doctor at least once a year; more frequently during very rapid growth, and discuss the need for dose changes. These drugs are given according to body weight; so, in growing children dose changes may be frequent.

If your child has a passion for sports, your child should be evaluated by a physician with expertise in LQTS to determine if continuation in competitive sports is reasonable.

Get additional medical advice if you are not comfortable with how things are going. Ideally, every family/patient with LQTS should be cared for by a heart rhythm specialist (pediatric or adult cardiac electrophysiologist) or even a long QT syndrome specialist. Do not hesitate to obtain a second opinion if you have any questions about your or your child’s treatment.

Learn CPR, encourage your child’s school district to implement an automatic external defibrillator (AED) in their schools, and consider obtaining a personal AED as part of the family’s LQTS safety gear. However, when well treated, remember that the AED should almost never be required.

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Thank you to Michael Ackerman, MD, Peter Schwartz, MD, and Arthur Wilde, MD for assistance and edits in this brochure.

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The QT interval is a time interval on the ECG. It represents the time from the electrical stimulation (depolarization) of the heart’s pumping chambers (ventricles), to the end of the electric refractory period, during which the heart muscle is refractory to a new stimulus. It is measured in milliseconds and closely approximates the time from the beginning of the ventricle’s contraction until the end of relaxation.

The QT interval varies in each person and between persons like most physiologic parameters, such as blood pressure. In particular, the QT varies with the heart rate. It shortens as the rate increases and lengthens as the rate decreases. Therefore, there is a range of normal or healthy QT intervals. But consistently, the long QT heart often recharges sluggishly or inefficiently as evidenced by a "prolonged QT interval" on the ECG.

To determine if a given QT is normal for a given heart rate, the QTc is corrected for the heart rate using a simple mathematical formula, and the resultant quantity is called the heart-rate corrected QT interval or the QTc. The QTc is the value that doctors generally use when assessing for LQTS. The average QTc value is around 400 +/- 20 ms. Probably less than 380 ms and 440 ms, which is the range doctors generally use when assessing for LQTS.

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Importantly, as an incidental ECG finding, QTc values exceeding these cut-off values do NOT equal the diagnosis of LQTS. However, not all LQTS patients have a prolonged QTc on the initial ECG. In fact, about 30 – 50% have a QTc that overlaps with the normal range and at least 25% of affected relatives have a QTc < 440 ms. QTc’s in this range are inconclusive, and therefore, the possibility of LQTS must be clarified by means other than ECGs. For example, determining the QTc in first degree relatives, regular or 12-lead 24 hour Holter monitoring, bicycle or treadmill stress testing, and genetic testing. If these additional tests are necessary to unmask or catch LQTS, all the tests must be interpreted carefully because of the potential for false positives.

How is the diagnosis made?

LQTS is suspected based upon the patient’s personal story, historical family history, and careful examination of the ECG. If the story is suspicious, a QTc exceeding 470 ms in males and 480 ms in females is sufficient evidence for a diagnosis of probable LQTS. Assuming that medications which prolong the QT interval or other QT-prolonging medical conditions have been ruled out Importantly, as an incidental ECG finding, QTc values exceeding these cut-off values do NOT equal the diagnosis of LQTS. On the other hand, a QTc of less than 400 ms in males and 410 ms in females makes the diagnosis unlikely. The computer generates formulas, but the final decision is made by the physician, who bases his or her decision on all of the above factors.

How is LQTS inherited, and who in a known or suspected family should be tested?

LQTS is usually inherited by autosomal dominant transmission. This means that it generally affects boys and girls equally, and that each child of an affected parent has a 50% chance of inheriting the genetic abnormality. In a really large family close to 50% of the children would inherit the LQTS-causing genetic abnormality. In average size families, it can range from all to none, and each child has a 50% chance (like a coin-toss) of inheriting the particular disease gene. Once a family member is identified with LQTS, it is extremely important that other family members be tested for the syndrome. It is especially important to know which parent and grandparent has the abnormality, since brothers and sisters, aunts, uncles, nephews, nieces, and cousins on the affected side are potentially at risk.

How should the diagnosis be suspected?

• In any young person with unexplained syncope (fainting), unexplained seizures, or unexplained cardiac arrest or sudden death.

Usually, a careful history of the events surrounding the syncope differentiates LQTS-induced syncope from the common faint, known as vasovagal or neurocardiogenic syncope. The LQTS faint is usually precipitated and without warning. It often occurs during or just after physical exertion (including swimming), emotional excitement including fear or sudden auditory arousal (such as a doorbell or alarm clock), but may occur during sleep or at rest. Conversely, in vasovagal syncope, most times there are warning symptoms, such as dizziness, blurring or blackening of vision, tingling or sweating, for seconds to even minutes prior to the syncope. Also, a precipitating event is usually present, commonly pain, injury, nausea, or an unpleasant or stressful experience.

• When there is a family history of unexplained syncope, unexplained seizures, or sudden death in young people. As noted above, about one in 200 people in the general population have LQTS.

What is the treatment and who should be treated?

All symptomatic patients should receive treatment. Most children and young adults who do not have symptoms of LQTS, if they do not experience symptoms. This is because symptoms might occur and sudden death can be the first symptom. At present, it is not possible to tell which child or youth is destined to have symptoms. Thus, preventive treatment is recommended for most. The usual treatment

This initial family relative screening, by ECG, is extremely important so that all affected family members are identified and treated early in order to prevent the tragic and unnecessary deaths that this disorder can cause. Importantly, once the diagnosis of LQTS has been established firmly in the family, there is no QTc value on the ECG that can be viewed as reassuring enough to dismiss the relative. If the genetic test is positive for the index case, then cascade genetic testing of the family members that yields a negative genetic test result AND a normal ECG can enable that family member to be dismissed as unaffected.