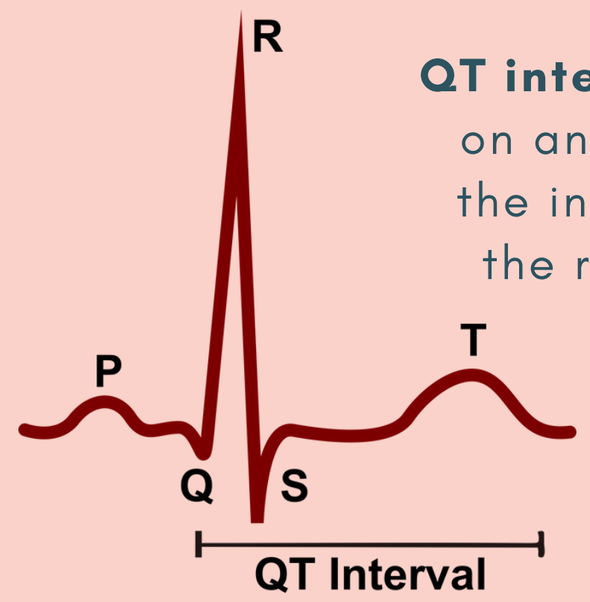
Long QT and Genetic Testing

Long QT syndrome (LQTS) is the most common inherited arrhythmia condition, affecting approximately 1 in 2,500 people around the world.

LQTS is a heart rhythm disorder that can cause fast, chaotic heartbeats that trigger sudden fainting spells, seizures, and even sudden death. It is a serious condition, but is treatable.



QT interval is measured on an ECG. The longer the interval, the higher the risk for symptoms.

Diagnosis and Treatment

- LQTS is usually a chronic condition that cannot be cured, but **can be managed**.
- Most patients are prescribed a medication known as a **betablocker**. For those few patients that still have symptoms despite medication, a surgical procedure called "left cardiac sympathetic denervation", or an implantable defibrillator may be recommended.
- **There can be several causes of a prolonged QT interval**, including genetic disorders, medical conditions and medication side effects. Genetic factors and environmental factors can play a role.

Genetic Testing

Genetic testing starts with an individual who has signs of LQTS. If a disease-causing variant is found, family members can be offered genetic testing.

3 possible results from genetic testing:

Positive

70-80% of the time, a disease-causing variant is found. This provides an explanation for why a person has LQTS. → Family members would have the option of genetic testing to find out if they are at risk for LQTS.

Negative

No disease-causing variant is found. This does not take away the clinical diagnosis of LQTS. LQTS may still be genetic, but the test did not find the cause. → We may recommend further genetic tests, and family members may need cardiac surveillance.

Indeterminate

We may find a variant of unclear significance, where we are not sure if it is the cause of LQTS. These patients should be referred back to Genetics every 5 years for updated information. → This result can't be used to test family members, so cardiac surveillance is required.

This reference is for educational purposes only.
If you have any questions, ask your health-care provider.

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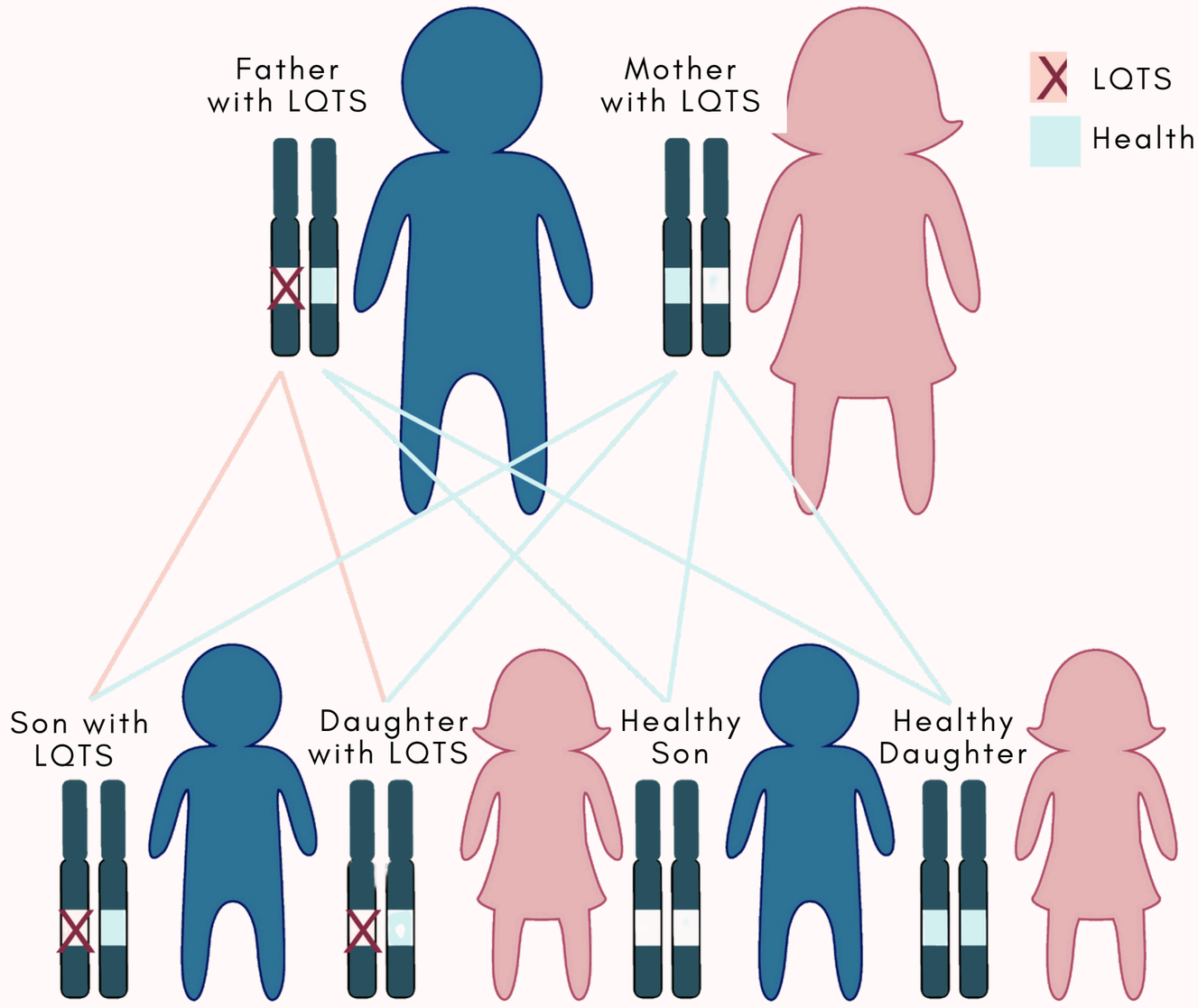

Inheritance

Usually autosomal dominant. Each person with LQTS has a 50% chance of passing on a genetic risk factor for LQTS to his or her children (both sons and daughters).

First-degree relatives of someone with LQTS (children, siblings and parents) should be evaluated for LQTS. The first test is typically an ECG, which needs to be reviewed by a specialist. An exercise treadmill test may also be recommended.

Genetic testing and Insurance

A law called the **Genetic Non-discrimination Act** prevents insurance companies from viewing or using the results of genetic testing to set prices or decide whether you get insurance. Provincial health insurance in Ontario and Quebec (OHIP and RAMQ) is not affected by your genetic results or by a medical diagnosis.

For private insurance (travel, life, disability etc.) while genetic test results are protected, other information about someone's current diagnoses, symptoms, and non-genetic results (e.g. cardiac screening) can be used by insurers when someone applies for a policy. Family medical history can also be used by insurers.

Resources: www.sads.ca | www.cagc-accg.ca to find a genetic clinic | www.cardiomyopathy.org www.ottawaheart.ca/heart-condition/arrhythmias-heart-rhythm-disorders www.ottawaCVgenetics.ca

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