Long QT Syndrome should be considered in any otherwise healthy child or young adult who:

- Experiences syncopal (loss of consciousness) or near syncopal spells principally during physical exertion or emotional stress - particularly in cases where the events are repetitive. *NOTE:* Episodes occurring during emotional stress are somewhat less specific because vasovagal (the common faint) episodes also may occur during emotional excitement. However, it's unusual for the common faint to occur during physical activity.
- Has a family history of unexplained sudden death in otherwise healthy young persons. *NOTE:* Some of the conditions which should be questioned are unexplained death during swimming, death during seizures, a family history of "seizure" disorders and other unexplained deaths.
- THE SCREENING PROCESS
- THE RESTING ELECTROCARDIOGRAM:
- This test will detect many individuals with the Long QT Syndrome. However, the QT interval is sometimes only moderately prolonged, and can be normal even in individuals with the Syndrome. Therefore, the resting electrocardiogram, while very helpful in many patients, is not perfect. Further information can be obtained by evaluating several electrocardiograms (ECG's), and ECG's on family members. Since this is a genetic condition in most cases, a child suspected of having the Long QT Syndrome would most likely have a parent with the syndrome, and very likely siblings with the syndrome also. Therefore, evaluation of the electrocardiograms of the parents and siblings of a suspected Long QT Syndrome patient can also assist in making the diagnosis.

THE EXERCISING ELECTROCARDIOGRAM:
QT intervals that are somewhat borderline or even normal at rest may become more distinctly abnormal during exercise, clarifying the diagnosis or clarifying that the patient does not have the Long QT Syndrome if the QT interval shortens. An exercise test which allows the individual to exercise for 10 or 15 minutes without achieving a heart rate more than 150-160 beats per minute works best for this process. The ECG's are then evaluated for the behavior of the QT interval at each stage of exercise and, particularly important, just after exercise in the early recovery. The QT interval is prolonged or even prolongs further during exercise and recovery in Long QT patients.

EPINEPHRINE CHALLENGE:
QT intervals that are somewhat borderline or even normal at rest may become more distinctly abnormal during infusion of a medication called epinephrine in a controlled and monitored manner, usually within a clinical electrophysiology laboratory.

GENETIC TESTING:
Clinical genetic testing is positive in about 70% of individuals considered to have long QT syndrome clinically. If positive, the specific type of long QT syndrome can be helpful in guiding restrictions and modifications to lifestyle, and predicting prognosis and response to interventions such as beta-blockers and pacing. A negative test does not rule out long QT syndrome in an isolated individual. When one member of a family is genetically identified with a Long QT

Syndrome, sequential genetic testing of first-degree relatives has a high accuracy in determining which other family members are also affected or unaffected. A professional with experience in genetic assessment and counseling should interpret long QT genetic testing. Clinical genetic testing is currently performed in laboratories outside of Canada, following case by case application and approval through provincial Ministries of Health.

Dr. Robert Hamilton, M.D., F.R.C.P. (C)
Electrophysiologist
The Hospital for Sick Children, Toronto

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