A Preventable Cause of Sudden Unexpected Death in Epilepsy (SUDEP) Finally is Established

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There are many potential reasons for sudden unexpected death in epilepsy (SUDEP). One very simple cause is an electrical abnormality in the heart. Like the abnormal brain waves that occur during seizures, an irregular heart beat (arrhythmia) may also come and go. In many cases, the abnormal heart beats are unpredictable, and people with the abnormality live long and healthy lives. While a simple “skipped heartbeat” is a normal occurrence, some patterns are uncommon, and for unknown reasons, the heart may simply stop, and not restart.

As there are many different types of seizures, there are also many different types of arrhythmias. Some irregularities in heart rhythm are benign. They can occur in perfectly healthy people at rest or are triggered by stress, too much caffeine, even excessive exercise. Some arrhythmias are more serious. One of the best known heart rate abnormalities is the Long QT Syndrome (LQTS). This is named after a characteristic electrical abnormality, prolongation of a QT interval, that can be detected in a routine measurement from an electrocardiogram (ECG, also sometimes abbreviated EKG). The LQT abnormality is commonly associated with fainting spells (syncope), but can also be a risk factor for sudden death.

The initial evaluation of a fainting spell usually involves an ECG (small surface electrodes are placed on your chest and extremities and a recording of the heart is made for several minutes) and a measurement of your blood pressure. If the ECG test shows a prolongation of a QT interval the physician may recommend anti-arrhythmic medications, pacemaker, or other interventions depending on the findings on the ECG and patient symptoms. Genetic testing can be helpful in pinpointing the mutation that underlies the long QT interval arrhythmia. However, it is important to remember that a negative genetic test does not rule out the long QT syndrome and that a positive genetic test confirms LQTS only if the ECG study is abnormal. Therefore, even though LQTS gene tests can confirm the risk for an electrical abnormality, they are not as sensitive as the ECG test itself.

Each year many young patients with epilepsy die suddenly. The molecular mechanisms behind this tragic end have been unknown. In 2009, an epilepsy research laboratory discovered that a gene responsible for the most common form of LQTS can not only trigger fainting spells, and put patients at risk of sudden death, but can also cause epilepsy (1). This had been long suspected, but never proved. This is an important finding, because it means that a seizure, and not just a fainting spell, could signify that you have a heart rhythm disturbance such as LQT and are at risk for sudden cardiac death. A cardiology laboratory has also verified that many episodes in individuals with confirmed LQTS are likely to have epileptic seizures rather than simply fainting spells (2). These scientific discoveries are very important because for the first time we can begin to take real steps to prevent at least one cause of SUDEP. Until very recently, people with seizure disorders did
not routinely receive an ECG test, because there was no firm evidence of a relationship between LQTS and epilepsy. This important new evidence means that one way to reduce the risk of SUDEP is to screen for heart rhythm disorders with an ECG. If an abnormality is found, the doctor will consult a cardiologist for the appropriate treatment.

- When a doctor first orders an EEG for a person with a seizure, they should also consider an ECG.
- If you have epilepsy and have never had an ECG, ask your doctor if you should have one. It could save your life.

References


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