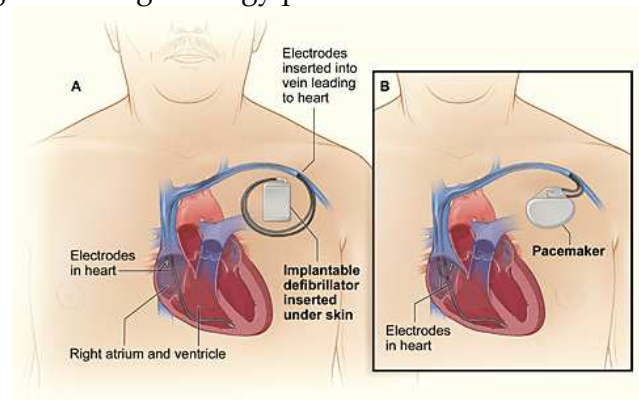


Long QT syndrome

Long QT syndrome is a rare heart rhythm disorder. It causes a very rapid heartbeat. These quick heart beats can trigger a sudden seizure or fainting spell. Sometimes, the heart beat so quickly for so long that it causes sudden death. Long QT syndrome is a rare disorder; however it is a severe, fatal illness. Symptoms usually appear during childhood and young adulthood.

There is no known cure for long QT syndrome; however, there are several treatments. Often, a combination of different treatments is used. One possible treatment option for long QT syndrome is an implantable cardioverter defibrillator. An implantable cardioverter defibrillator (ICD) is a small device that is used to help treat irregular heartbeats, in this case, those caused by long QT syndrome. An ICD is placed in the chest or abdomen. It uses electrical pulses or shocks to control arrhythmias (irregular heartbeats). ICD's are especially used in cases where arrhythmias are life-threatening and may cause SCA (sudden cardiac arrest). SCA can happen in those with long QT syndrome and most often causes death. The heart has an electrical system which naturally controls rate and rhythm of the heartbeats. With every heartbeat, an electrical signal is spread from the top of the heart to the bottom. When the signal travels it causes the heart to contract and pump blood around the body. When there is a problem with the heart's electrical system it causes an arrhythmia, like in long QT syndrome. Implantable cardioverter defibrillators use electrical shocks to treat the potentially fatal arrhythmias that occur in the ventricles of the heart. People with long QT syndrome often faint because ventricular arrhythmias mean that the heart cannot pump blood well. The electric shocks treat the arrhythmias and prevent death. The treatment by the electrical shocks sent to the heart is called defibrillation (and a device that defibrillates is called a defibrillator). ICD's monitor heart rhythm so that they can send electrical shocks if an irregular rhythm is detected in the ventricles so that a normal rhythm can be restored. Usually, low-energy impulses are used for defibrillation but sometimes these aren't enough to restore the normal heart rhythm. In this case, the ICD switches to high-energy pulses to defibrillate. It will also change to the high-energy pulses if the ventricles start to shake instead of strongly contracting. The shocks the ICD sends out are usually very effective in restoring the normal heart rhythm; however the high-energy pulses can be very painful. The ICD is implanted. Usually, the electrode wires pass through a vein to the right chambers of



the heart. The wires are then lodged in the apex of the right ventricle. Although ICDs may seem similar to pacemakers, they are actually quite different. The diagram to the left shows an IDC (A) and a pacemaker (B). The main difference between an IDC and a pacemaker is that a pacemaker is used to consistently correct bradycardia (slow heartbeat) whilst an ICD is used to treat sudden, life-threatening arrhythmia which may cause sudden death.

There are many benefits of ICDs. The main benefit is that the operation to implant the ICD is not very risky. The risk of having a complication such as bleeding, a punctured lung or an infection is only around 2% to 3%. Furthermore, the risk of death from this procedure is less than 1%. Life with an ICD is generally good. People with an ICD can live life to the fullest without making any major lifestyle changes.

There are some limitations of ICDs. Most physical activities can be performed by those with ICDs. However, great care must be taken to make sure that no excessive strain is placed on the arms, shoulder or torso. This is because this is around the area which the ICD is in. Putting excessive strain on any of these areas could damage either the electrodes going to the patient's heart or the device itself. In addition, those with ICDs should also avoid contact sports like American football, boxing, wrestling or contact karate. This is because hitting the chest area can damage either the electrodes or the ICD itself. Although most electronic equipment is safe to use with ICDs, there are some limitations on the use of electronic equipment. Most electronic equipment (for example, mobile phones) should be kept *at least* 15cm away from the ICD. Those with ICDs must avoid electronic equipment in a professional environment as well as equipment that use big magnets or that generates magnetic fields. MRI scanners can also interfere with how the ICD works. This means that those with ICDs cannot have MRI scans. If an MRI scan is needed for any reason, a patient with an implanted ICD should have a CAT scan instead. CAT scans are a safer alternative as they do not generate the magnetic fields that MRI scans generate. If external defibrillation is ever needed (for example from an AED (automated external defibrillator)), the paddles of the AED should not be placed directly on the device. This is because the shocks from the AED may damage the ICD.

Generally, an ICD is a very effective device. On average, those who receive an ICD live for two to four months longer than those who did not have an ICD. One out of every six patients lives two to three years longer than those who did not have an ICD. These statistics show that ICDs improve the life of people with heart disease,

like those with long QT syndrome. ICDs are very effective in stopping sudden death (SCA, sudden cardiac arrest).

Ethical factors hinder the use of ICDs. Some people think that it is not ethical to defibrillate. Defibrillation can save a patient's life. Without the shocks an ICD gives most people with long QT syndrome and other heart diseases could not survive. Long QT syndrome causes a rapid, uncontrolled heart rhythm which, if it last for long enough, can cause SCA, which in turn causes death. It is not possible to survive SCA without defibrillation. ICD shocks are painful; so many people don't want to use ICDs at the end of someone's life. However, if the patient is terminally ill the ICD may be deactivated to reduce the suffering of a patient. If however, the patient does go into a rapid, controlled heart rhythm, the ICD will not shock the patient and they will die unless they quickly receive other treatment, such as shocks from an AED (automated electronic defibrillator).

There are social factors that both help and hinder the use of ICDs. Long QT syndrome affects mainly children, and children with ICDs in them may encounter some social problems in their lives. Because of the limitations placed on those with ICDs, children cannot act like their peers and they cannot participate in all the activities their peers participate in. For example, a child with an ICD may be excluded because they cannot play certain sports, such as American football. Other treatments for ICD, such as beta-blockers (medicine) pose fewer restrictions on a child's lifestyle, however for a child that is at risk of sudden death from long QT syndrome, an ICD is often necessary. However, social factors also help the use of ICDs. Children that have severe long QT syndrome and do not have an ICD will need to have far more activity restrictions.

Generally, ICDs are a good treatment for long QT syndrome and they are effective in helping treat it.

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