**Condition:** Long QT syndrome

**Inheritance:**
Genetically heterogeneous.

**Genetic etiology:**
Most are due to mutation in various genes encoding sodium and potassium ion channels. Romano Ward syndrome (LQT 1-6) due to mutations in $KCNQ1$, $KCNH2$, $SCN5A$, $KCNE1$, $KCNE2$, encoding potassium or sodium channels. Jervell and Lange-Nielsen syndrome due to mutation in $KCNQ1$ (LQT1) or $KCNE1$ (LQT2), both potassium channels. Andersen-Tawil syndrome due to mutations in $KCNJ2$ encoding inward rectifier potassium channel.

**Frequency:**
Romano-Ward: 1/7,000; Jervell and Lange-Nielsen: 1.6 – 6/million; Andersen-Tawil syndrome: rare.

**Clinical features:**
Long QT syndrome is a disorder in which there is prolonged ventricular repolarization, leading to cardiac arrhythmia. There are three major syndromes, each of which is itself genetically heterogeneous. Romano-Ward is isolated long QT and is dominantly inherited. Jervell and Lange-Nielsen syndrome is long QT with sensorineural deafness and is autosomal recessive. Andersen-Tawil syndrome includes long QT, dysmorphic features, and periodic muscle weakness and is autosomal dominant.

**Management:**
Treatment with beta blocker medication or insertion of cardiac pacemaker.

**Genetic counseling:**
Based on establishing correct diagnosis; genetic testing available for some of the responsible genes. Individuals at risk should be carefully evaluated, since sudden death is possible in untreated individuals.