This presentation will focus with the management of the Long QT Syndrome (LQTS). The topics to be discussed comprise diagnostic criteria, importance and implications of genetic testing, and all major therapeutic options including gene-specific management. Diagnostic criteria have recently acquired a special role particularly for determining which patients should undergo genetic testing. The latter is important for allowing gene-specific strategies and for the identification of silent mutation carriers. Not to perform genetic testing in a clearly affected patient may carry medico-legal implications. The efficacy of beta-blocker therapy will be discussed in relation to non-genotyped patients and within the 3 major genetic subgroups with special attention for LQT3 patients who should not be denied the protection conferred by beta-blockers. Clinically relevant differences between different beta-blockers will be discussed. The efficacy and indications for Left Cardiac Sympathetic Denervation will be reviewed. Special attention will be devoted to correct and incorrect indications for the use of Implantable Cardioverter-Defibrillators (ICDs). Special emphasis will be paid to the use of a risk score, easily available in a medical office during a first visit, to assess the probability that ICD implant will be accompanied by appropriate shocks. Careful use of this score will help to avoid unnecessary ICD implants in this young population.

Keywords: Long QT syndrome, beta-blockers, genetics