Abstract

The mind-boggling progress in the understanding of the molecular mechanisms underlying the long QT syndrome (LQTS) has been the subject of many articles and reviews. Still, when it comes to the management of the patients affected by this life-threatening disorder, too many errors still take place, both in the diagnostic process and in the therapeutic choices. The price of these errors is paid by the patients and their families. This review is not directed to the relatively small number of LQTS experts who know what to do. It does not deal with genetics, with epidemiology, or with the well-known clinical manifestations. We have focused solely on the approach to diagnosis and therapy and we have directed this review to the average clinical cardiologist who, in his/her practice, sees occasionally patients affected or suspected to be affected by LQTS; the cardiologist who may know enough to manage them but not enough to be completely confident on his/her most critical choices. We have provided our personal views without making any attempt to blend differences whenever present. On most issues we agree fully but where we do not, we make it clear to the reader by indicating who is thinking what. The result may be unconventional, but it mirrors the challenges, often severe, that we all face in managing and protecting these patients from sudden death while also helping them live and thrive despite their diagnosis. We trust that this unabashed presentation of our clinical approach will be useful for both cardiologists and patients.

Keywords: Long QT syndrome, Sudden cardiac death, Left cardiac sympathetic denervation, Implantable cardioverter defibrillator, QT interval, Genetics

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For neonatal ECG screening there is no reason to consider long QT syndrome

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