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# Recent Advances in Congenital Long QT Syndrome

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## Commentary

During my residency, my program had initiated an inherited arrhythmia clinic under a pediatric electro physiologist. There is a similar clinic in my fellowship program and it might not be far-fetched to imagine that such inherited arrhythmia clinics will be a part of every child's hospital in the near future. The past decade has been an era of tremendous development in inherited arrhythmias and congenital Long QT Syndrome (LQTS) has been a poster child for this field. Given that there are numerous research articles in this field, I felt that a summary of recent advances in LQTS will be of benefit for fellows who might be involved in the care of these patients and in generating research hypothesis.

Understanding the underlying cellular mechanism of sudden cardiac arrest holds the key to developing therapeutic strategies for patients with LQTS. It is well known that patients with LQTS 1 (most common form of LQTS), due to KCNQ1 (delayed K+ rectifier channel) are at the highest risk for Sudden Cardiac Arrest (SCA)/Sudden Cardiac Death (SCD). Recent evidence points that specific mutations in KCNQ1 completely blunts the affected channel's responsiveness to exercise induced protein kinase A phosphorylation leading to further prolongation of QT interval [1]. New evidence have also shown that LQTS patients have localized regions of action potential duration that causes steep depolarization dispersion on non-invasive Electro Cardio Graphic mapping Techniques (ECGI) [2]. This demonstrated a substrate for arrhythmogenesis, which was not shown by surface ECG and could be used for risk-stratification.

One of the major breakthroughs in pediatric cardiology has been the study of congenital heart disease in the fetus, which has also permeated to LQTS. Fetal rhythm has been traditionally evaluated by using 2D, M-Mode or pulse Doppler during fetal echocardiography. Fetal Magneto Cardio Graphy (fMCG), a new investigative modality uses magnetometers that can non-invasively assess the electromagnetic conduction of the fetal heart. fMCG has been used to not only diagnose fetuses with LQTS but also identify those who are high-risk [3]. No specific etiology is found in upto 60% of fetal demise, new research now has linked dysfunctional LQTS-associated ion-channels in vitro among 8.8% of these deaths [4].

Although there have not been new therapeutic strategies of LQTS, there have been research on streamlining existing modalities. A recent study in addition to confirming that nadolol is one of the most beta blocker to use in LQTS2, has demonstrated that propranolol a commonly used beta-blocker in the setting of LQTS, and was the least effective among this class of drugs for high-risk group. The accompanying editorial suggested further research is needed to confirm or refute these findings prior to changing clinical practice [5]. Another pertinent study suggests that a cohort of children with LQTS who have QTc interval persistently <470 msec, who do not indulge in high risk activities and do not have certain high risk mutation might not need to take beta-blockers [6].

Recently formulated expert consensus recommendations on risk-stratification and management are helpful guide to the management of these patients. While Beta-blockers are the main-stay recommended therapy, implantable cardioverter defibrillator and left cardiac sympathetic denervation are recommended in selected high-risk patients [7]. Diagnosis of LQTS is devastating news for competitive athletes; however recent statement from ACC/AHA may allow certain individual to still participate. Asymptomatic athletes who were genotype positive/phenotype positive and symptomatic athletes with LQTS or manifest LQTS could also be considered to return to sports if they are asymptomatic after institution of treatment along with precautionary measures [8].

With wide spreading awareness of prolonged QT interval and congenital LQTS among both the medical community and the general population, keeping pace with recent advances is a necessity for pediatric cardiology FITs. Although LQTS might not be a commonly encountered diagnosis in daily practice, inadequate management of this condition may be severe consequences in the form of SCD. Being up-to-date on current guidelines is an easy

way of keeping pace with evidence based therapeutic strategies.

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