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Wearable cardioverter defibrillators for patients with long QT syndrome

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ABSTRACT

Background: Long QT syndrome (LQTS) is a potentially lethal cardiac channelopathy, but with the appropriate treatment strategy, such as beta-blockers, left cardiac sympathetic denervation (LCSD), and/or an implantable cardioverter defibrillator (ICD), most LQTS-triggered tragedies can be avoided. Since 2001, wearable cardioverter defibrillators (WCD:LifeVest™) have been available clinically.
Objective: Herein, we evaluated the use and outcome of WCDs in patients with LQTS.
Methods: We performed a retrospective review of 1027 patients with LQTS to identify patients who received a WCD, and collected pertinent clinical information regarding their LQTS diagnosis as well as indication and experience regarding use of the WCD.
Results: Overall, 10 LQTS patients (1%, 8 females, age at diagnosis 29 ± 18 years, mean QTc 488 ± 34 ms) were prescribed a WCD. Most common indication for WCD was as bridge to treatment during (temporary) situation of assessed high risk of sudden cardiac arrest (SCA; $n = 6$). The mean time of WCD use was 24 days (range 0 to 114 days). One patient (female, age 42, LQT2) received an appropriate VF-terminating shock 2 days after receiving her WCD. No inappropriate treatments or adverse events from wearing the WCD have occurred.
Conclusions: A WCD can be considered in patients with LQTS deemed to be at high risk for SCA while up-titrating beta blockers, considering ICD therapy, or when navigating short term periods of increased SCA-risk, like the post-partum period in LQT2 women, ICD revision or temporary inactivation, or during short term administration of known QT prolonging medications.

1. Introduction

Congenital long QT syndrome (LQTS) is a highly treatable yet still potentially lethal cardiac channelopathy. LQTS affects approximately 1 in 2000 people [1] and is a heritable cardiac disease whereby patients are at an increased risk for LQTS-triggered syncope, seizures, and sudden cardiac arrest (SCA) following the characteristic ventricular arrhythmia of torsades de pointes [2]. Although 17 LQTS-susceptibility genes have been identified, approximately 75% of LQTS can be explained by mutations in three genes (*KCNQ1*, *KCNH2*, and *SCN5A*) that encode pore-forming subunits of ion channels (Kv7.1, Kv11.1, and NaV1.5 respectively [3]. While each of the major LQTS subtypes has its unique pro-arrhythmic triggers, outside factors may also contribute to the risk level of an individual having a LQTS-triggered cardiac event.

Aside from our standard, LQTS-directed preventative/safety measures (avoidance of QT prolonging drugs, daily fish oils supplement, advice for proper hydration and fever reduction, and purchasing an automated external defibrillator (AED)), therapeutic options for LQTS include pharmacotherapy (principally beta-blockers),surgical therapy (principally left cardiac sympathetic denervation (LCSD)), and device therapy (primarily an implantable cardioverter defibrillator (ICD)).

Since 2001, wearable cardioverter defibrillators (WCD; LifeVest™) have been available clinically and, although used primarily in adults with a recent myocardial infarction, could provide an option for patients with LQTS [4]. Not meant to be a permanent solution, WCDs can provide temporary protection during the evaluation period during periods of heightened risk for SCA or serve as a bridge to an ICD. Herein, we evaluated the use and outcome of WCDs in patients with LQTS.

2. Methods

For this IRB-approved, retrospective study, we reviewed the electronic medical records of 1027 adult and pediatric LQTS patients evaluated at Mayo Clinic's Long QT Syndrome/Genetic Heart Rhythm Clinic between 2000 and 2017 to identify use of WCD. For patients who were prescribed a WCD, pertinent clinical information regarding their LQTS diagnosis as well as indication and experience regarding use of the WCD was collected, including but not limited to LQTS genotype, QTc measurement, symptomatology, family history of LQTS and SCA, duration of WCD use, complications, appropriate or

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inappropriate shocks, and additional outcomes. Overall utility of the WCD in this patient cohort as well as detailed experiences and outcome of patients prescribed the wearable device were evaluated and described.

3. Results

Overall, ten patients (10/1027; 1%) were prescribed a WCD between 2000 and 2017 (nine adults and one pediatric patient). Demographics of these patients are summarized in Table 1. In brief, there were ten patients (eight females (80%)), with a mean age at LQTS diagnosis of 29 ± 18 years (range 10–66 years) and mean age at WCD of 34 ± 16 years (range 15–66 years). Baseline QTc was 488 ± 34 (438–543 ms). The genotypes included four patients with LQT1, four with LQT2, one with compound LQT2, and one with LQT3. Six patients (60%) had a family history of LQTS and six patients (60%) had a positive family history for SCA. The mean duration of WCD use was 24 days (range 0 to 114). One patient (**Case 3**; discussed below) ultimately chose not to wear her device explaining the low end of the range.

Seven of the 10 patients (70%) were previously symptomatic, and documented symptoms included syncope, TdP and out of hospital cardiac arrest with VF-terminating shock from an automatic external defibrillator (AED). Clinical and WCD-related details for each of these ten patients are summarized in Table 2.

The most common indication for use of the WCD was as a bridge to treatment during initial diagnostic work up and protection from SCA while either achieving a therapeutic dose of beta blocker – generally nadolol at 1–1.5 mg/kg/day [5–7] – or facilitating surgical date for LCSD surgery in six of ten patients (60%; **Cases 1**, **2**, **4**, **5**, **7** and **9**). For two of these six patients (**Cases 1** and **7**), this was compounded by an additional diagnosis, unrelated to LQTS, that required prescription of medications known to prolong the QT interval (levofloxacin and compazine respectively). Additionally, these patients were considered to be at temporary high risk due to frequent electrolyte imbalance and potential for febrile episodes.

For the three remaining patients (**Cases 6**, **8**, **10**), the WCD was prescribed during a time of ICD malfunction. **Patient 6** presented to his local cardiology clinic with a fractured ICD lead. His ICD was implanted in 2006 and he had remained asymptomatic from his LQT2 substrate since. The patient's ICD was deactivated due to risk of inappropriate shock from the fractured lead and he was outfitted with a WCD. He requested that the ICD revision be done at Mayo Clinic and due to long distance travel and scheduling conflicts for the patient, the WCD was prescribed as a bridge until his ICD lead revision. For **patient 8**, the LQT3 SCA-prevention program had been ICD solo therapy for the past 15 years during which she had been symptom free. She did however have two inappropriate ICD shocks due to a fractured lead for which lead revision was required. The patient was transferred to Mayo Clinic from her local emergency room after being fit for a WCD. **Case 10** presented to her local cardiology clinic with an ICD lead fracture. This asymptomatic LQT2 female received her prophylactic ICD in 2004 and has not received an ICD shock. She was prescribed a WCD after her ICD was deactivated because of the risk of inappropriate shocks from the fractured ICD lead. The patient requested ICD lead revision at the Mayo Clinic and wore the WCD while this was facilitated. Herein, the WCD provided SCA protection while the ICD was deactivated and a surgical date for ICD replacement was facilitated in these three patients.

In one young female patient (**Case 4**), the WCD was used for SCA prevention after she experienced a swimming-associated cardiac arrest. The patient and her family did not want to proceed directly to an ICD. A WCD was prescribed and the patient was referred to Mayo Clinic. Ultimately, she was diagnosed with LQT1, which was confirmed by genetic testing. The patient had minimally invasive LCSD surgery and was started on nadolol. She has not had any LQT1-triggered symptoms following LCSD and initiation of beta blocker therapy.

During the time of WCD use, one patient (10%) received an appropriate VF-terminating shock from her WCD (**Case 5**). This patient was a 42-year-old female with compound LQT2 secondary to variants in *KCNH2* (V603I and L799sp) who came to Mayo Clinic seeking a second opinion and review of her current LQT2-directed treatment program. Previously, she had experienced seven to eight seizures or seizure-like episodes since the age of 20. She presented at age 42 to her local health care provider with palpitations and was evaluated subsequently by a cardiologist, where she was told she had a prolonged QT interval on her ECG with a recorded QTc of 543 ms. However, at that time, her local cardiologist did not make a definitive diagnosis or start treatment. The next day, she had a syncopal, seizure-like event at home, followed by another syncopal episode at her clinic appointment three days later. A Holter monitor was placed and a cardiologist notified. Because of the sequelae of events, the referring cardiologist became suspicious of LQTS and referred her to Mayo Clinic. Following phone consultation with our team, the cardiologist dismissed her with a WCD, but against our advice, did not initiate beta blocker therapy. Two days later, during an emotional moment hugging a family member, she lost consciousness. The WCD captured the event and appropriately discharged a VF-terminating shock (Fig. 1). An ICD was placed two days later and beta blocker therapy was initiated. In the four years since this event, she has had one appropriate, VF-terminating ICD shock during a period of postoperative vomiting and electrolyte imbalance.

None of the ten patients with WCD received any inappropriate shocks from the vest. Other complications or side effects associated with WCD-use were related to annoyance of false alarms during (extended) time of wearing the device in two of ten patients (**Cases 1** and **6**). One patient (**Case 6**) removed the WCD during the night of one of the five days he was wearing it because of several episodes of false alarms caused by poor contact for ECG analysis. The other patient (**Case 1**), who wore the WCD for 70 days, complained that the straps became loose after extended wear causing false alarms due to poor connections. In addition, another patient (**Case 9**) explained that she lived in a warm and humid climate and the WCD made her feel overheated, but she continued to wear the WCD. Overall in this small cohort, the WCD was reasonably tolerated, with patients wearing the device for most of the day, only to take it off for changing clothes or showers.

Nevertheless, one patient (**Case 3**) included in this cohort chose not to wear the device. This 28-year-old female with LQT1 had a history of syncope and had an ICD placed at the time of her diagnosis. She received multiple inappropriate ICD shocks and came to Mayo Clinic seeking additional treatment options. Her ICD was deactivated and beta blocker therapy treatment was started. The patient returned to Mayo Clinic after becoming pregnant. At that time, because of the family's personal uncertainty as to how the pregnancy would be tolerated as well as a fear of a LQT1-related event to the mother and risk associated to the unborn child, the patient and her family requested an additional safety measure. Despite an informed discussion of the risk of post-partum events being associated particularly with LQT2 and not – as in this patient – LQT1, she requested a WCD and was provided a prescription for the device. However, in the end, she decided not to wear the device, possibly because

Table 1
Demographics of patients with WCD.

	Cohort ($N = 10$)
Sex (male/female)	2/8
Mean age at diagnosis yrs. (range)	29 ± 18 (10–66)
Age at WCD yrs. (range)	34 ± 16 (15–66)
Baseline QTc ms (range)	488 ± 34 (438–543)
Symptomatic prior to diagnosis n (%)	7 (70)
Family history of LQTS n (%)	6 (60)
Family history of SCA n (%)	6 (60)
Duration of WCD Use	0 days to 114 days
Genotype positive n (%)	10 (100)
LQT1	4 (40)
LQT2	4 (40)
LQT2 compound	1 (10)
LQT3	1 (10)