# Challenges of ICD Therapy in the Management of Long QT Syndrome

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## Introduction:

There is an increasing recognition of the subtle manifestations of the various inherited myocardial channelopathies and their life threatening consequence of sudden cardiac death in children, adolescents and young adults. Since the cause of death is polymorphic ventricular tachycardia degenerating to ventricular fibrillation, there has been a concomitant increase in the use of implantable cardioverter-defibrillators in these patients. The limitations associated with current ICD therapy combined with some unique and bizarre T wave abnormalities in patients with the Long QT Syndrome[1] may combine to cause T wave oversensing and delivery of inappropriate therapy in the setting of normal sinus rhythm. This case report describes one such patient and the manner in which this problem was managed.

## Case Report:

The patient is a 38 year old woman who had been diagnosed as having epilepsy since childhood. She presented with syncopal spells that degenerated into epileptic seizures. EEG studies failed to demonstrate an epilepitic focus and a multiplicity of anti-epileptic agents failed to prevent her recurrent syncopal spells and seizures[2,3,4]. When her father and another sibling were diagnosed with the Long QT Syndrome (and an ICD implanted in her father), she was referred for evaluation. A diagnosis of LQT3 was made based on genetic testing (SCN5A defect) and she underwent an uncomplicated implantation of a dual chamber ICD (St. Jude Medical Epic™ + DR V-236).

On the first post-operative day, she had three inappropriate shocks due to T wave oversensing in the setting of a sinus rate of 110 bpm and the implanted system double counting both the QRS and T wave for an effective rate of about 220 ppm. The peak of the T wave was detected 280 to 290 ms after the intrinsic deflection of the QRS complex (Figure 1). There was single zone therapy with the VF zone starting at 200 ppm (300 ms). The programmed ventricular sensitivity was 0.3 mV. This is the nominal value with the least sensitive setting that is allowed being 1.0 mV.

The Auto Sensitivity Control algorithm in the ICD was reprogrammed to eliminate the T wave oversensing by increasing the ventricular post-sense Threshold Start from 50% to 62.5% (of the peak QRS amplitude) and ventricular post-sense Decay Delay increased from 0 to 190 ms. Following this, repeat DFT testing was performed to confirm that there was no adverse impact of these new settings on the recognitiion of VF. At the same time, the sensitivity remained at 0.3 mV to maximized detection of VF episodes.

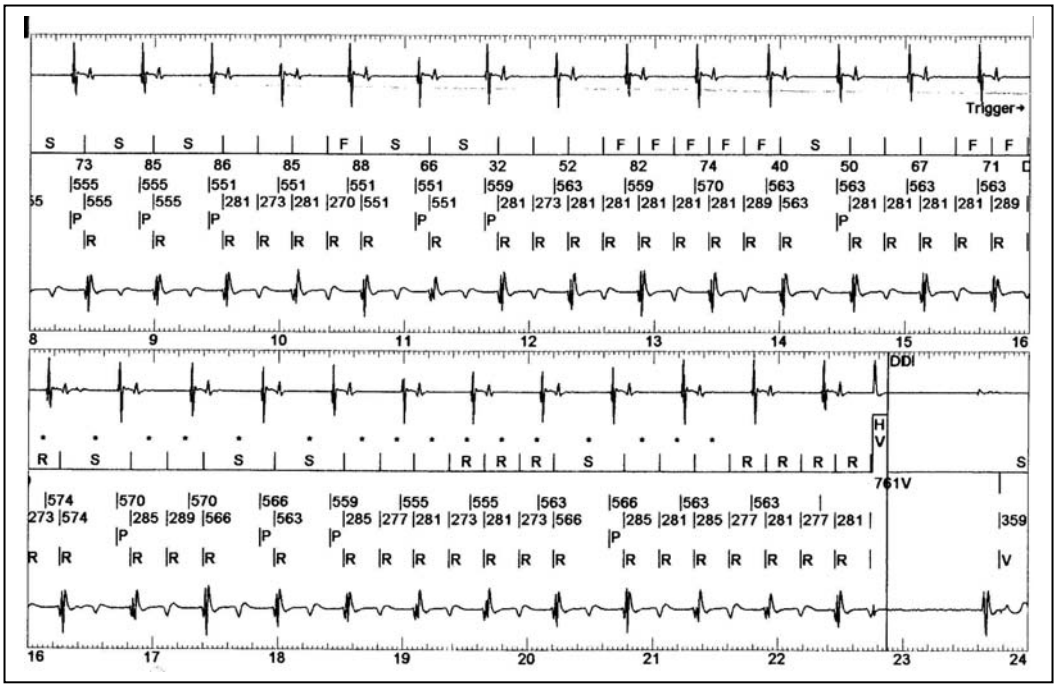


Figure 1: Two sections of the stored EGM from one of the inappropriate therapies associated with T wave oversensing. The top EGM in each section is the atrial EGM, and the bottom EGM in each section is the ventricular EGM. The “F” markers above the solid line with up-tick markers indicate that the detected rate fell in the fibrillation zone. The “S” markers above the solid line indicate that the rate was below the tachycardia rates and is labeled sinus but this must occur for 5 consecutive cycles for the device to declare the tachycardia episode ended and thus suspend therapy. The “R” markers above the solid line indicate that the tachyarrhythmia was reconfirmed during capacitor charging until the moment at which a high voltage (HV) shock was delivered.
“R” and “P” with side tick markers above the ventricular EGMs identify the detected events as native R or P waves.

Since these programming changes were implemented, there have been no further inappropriate therapies delivered. Since implant, she has not had any further syncopal episodes. .

**Discussion:**

To optimize detection of ventricular fibrillation in the shortest possible time, virtually all ICDs utilize relatively broad band-pass filters in their sensing circuit, maintain a sensitivity setting that is usually more sensitive than the least sensitive setting of a standard pacemaker and have a very short ventricular refractory period. The range of programmable maximum sensitivity settings in the Epic + DR ICD is from 0.2 mV to 1.0 mV with a nominal value of 0.3 mV. The normal T wave is predominantly comprised of low frequency signals. These would be effectively eliminated by the sensing circuit of a standard pacemaker. The broad band-pass filter of the ICD sensing circuit does not eliminate all of these signals as fibrillatory signals are often in this range predisposing ICDs to T wave oversensing. Another option available in a pacemaker is a programmable ventricular refractory period extending out as far as 300 or even 400 ms but this is not an option in an ICD as a long refractory period would preclude recognition of VF. While the ventricular refractory period in the Epic+DR ICD is programmable, it has only two options (125 ms and 157 ms), neither of which would be sufficient to preclude T wave oversensing.

The Epic family of ICDs utilizes an Auto Sensitvity Control algorithm. Despite the standard sensing circuit being refractory upon a sensed event, the ICD continues to analyze the native QRS and identifies the peak signal amplitude. Upon the end of the refractory period, the sensitivity is automatically adjusted to to a programmable percentage (in this case, an initial nominal value of 50%) of the peak amplitude. This is termed Threshold Start. The sensitivity then proceeds to become progressively more sensitive until another event is sensed, or the programmed maximum sensitivity value is reached. If it reaches the programmed maximum sensitivity level, it remains there until an R wave is detected or the basic escape interval is completed and a pacing stimulus is delivered. The point at which the system starts to become more sensitive is termed Decay Delay. A Decay Delay value of 0 ms means that the increase in sensitivity starts immediately. The start can be delayed up to 220 ms. The stored EGMs retrieved from the inappropriate therapy episodes can be sent to St. Jude Medical's Tachycardia Technical Service section and using a Ventricular Post-Sensed Oversensing Calculator (Figure 2), the amplitude of the inappropriate signal measured and recommendations provided for programming the Threshold Start and Decay Delay. In this case, Threshold Start was increased to 62.5% of the baseline value and Decay Delay was extended to 190 ms. The measured T wave amplitude was 1.278 mV. By setting the above values, the sensitivity of the device at the timing of the prior T wave detection would be 1.764 mV effectively precluding T wave detection.

**Summary:**

A young woman was misdiagnosed as having epilepsy until a diagnosis of Long QT Syndrome was suspected after family members were diagnosed with this condition. She subsequently underwent implantation of a dual chamber ICD but this therapy was compromised by T wave oversensing associated with the bizarre and large T waves that are associated with this syndrome. Some unique capabilities of the Auto Sensitivity Control algorithm in St. Jude Medical's Epic+DR ICD system allowed the device to be programmed so as to preclude the T wave oversensing while maintaining the high sensitivity and short refractory periods needed to optimize detection of ventricular fibrillation and polymorphic ventricular tachycardia.

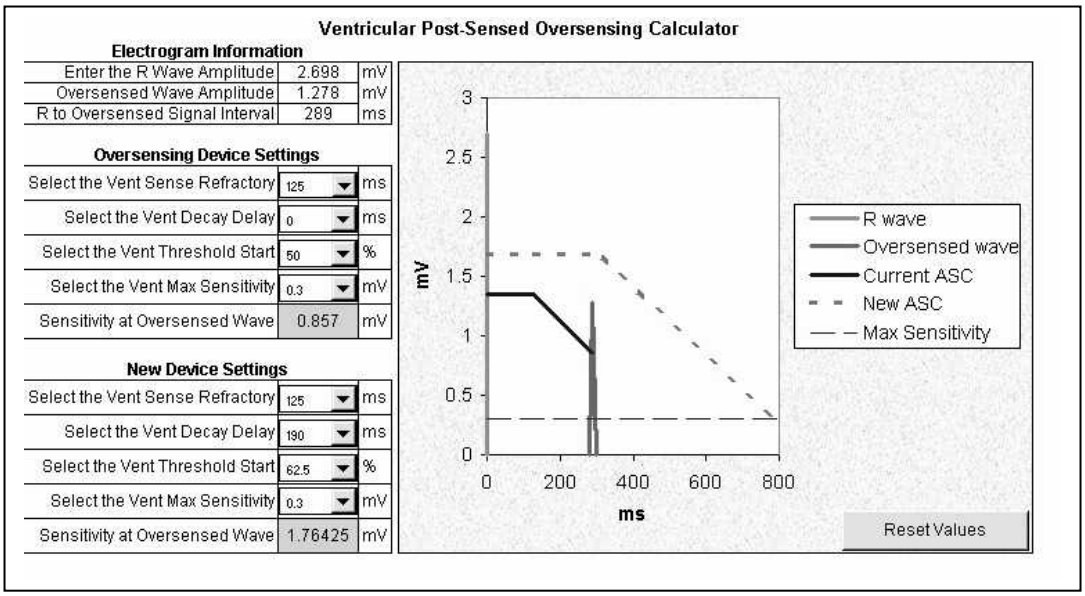


Figure 2: The graphic display of the Ventricular Post-Sensed Oversensing Calculator. The solid lines represent the original programmed parameters, the specific values being displayed in the original device settings in the middle box. The modeled parameters and results are shown by the dotted line labeled New ASC with the specific calculated measurements shown in the lower box to the left of the graphic display.

## Bibliography:


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