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A Young Patient with Dilated Cardiomyopathy and Out-of-Hospital Sudden Death: From Randomized Trials and Guidelines to Real Life Patients.
Part III: What went wrong?

Sami Viskin, M.D.
Department of Cardiology, Tel Aviv Sourasky-Medical Center and Sackler School of Medicine, Tel Aviv University, Israel.

Address for correspondence:

Sami Viskin, M.D.

Department of Cardiology

Tel Aviv Medical Center

Weizman 6

Tel Aviv 64239

Israel

Tel + Fax: 972-3-6974416

E-mail: saviskin@tasmc.health.gov.il

Part III: What went wrong?

The patient was found dead by a relative, still seated in front of the television. The television was still on. The telephone records showed that he did not call for help. Therefore, the death was presumed to be sudden, without premonitory symptoms, and most consistent with an arrhythmic mechanism. Though autopsy was not done, we performed a post-mortem interrogation of the implanted ICD. The device memory showed that the patient was free of arrhythmias until the day he died. The retrieved electrograms, stored in the device's memory at the presumed time of death, are shown in figures 2-7.

Figure interpretation. The stored electrograms show the cause of death. All the traces are from the same event but the traces are not always continuous because the arrhythmic event was very long so only the essential traces are shown.

The initial arrhythmia (figure 2) is atrial flutter with atrio-ventricular (AV) block and appropriate ventricular pacing at the programmed lower rate limit, which is 40 beats/min (pacing interval =1500 msec). Then, a rapid polymorphic VT is appropriately detected by the ICD in the programmed VF zone (figure 2).

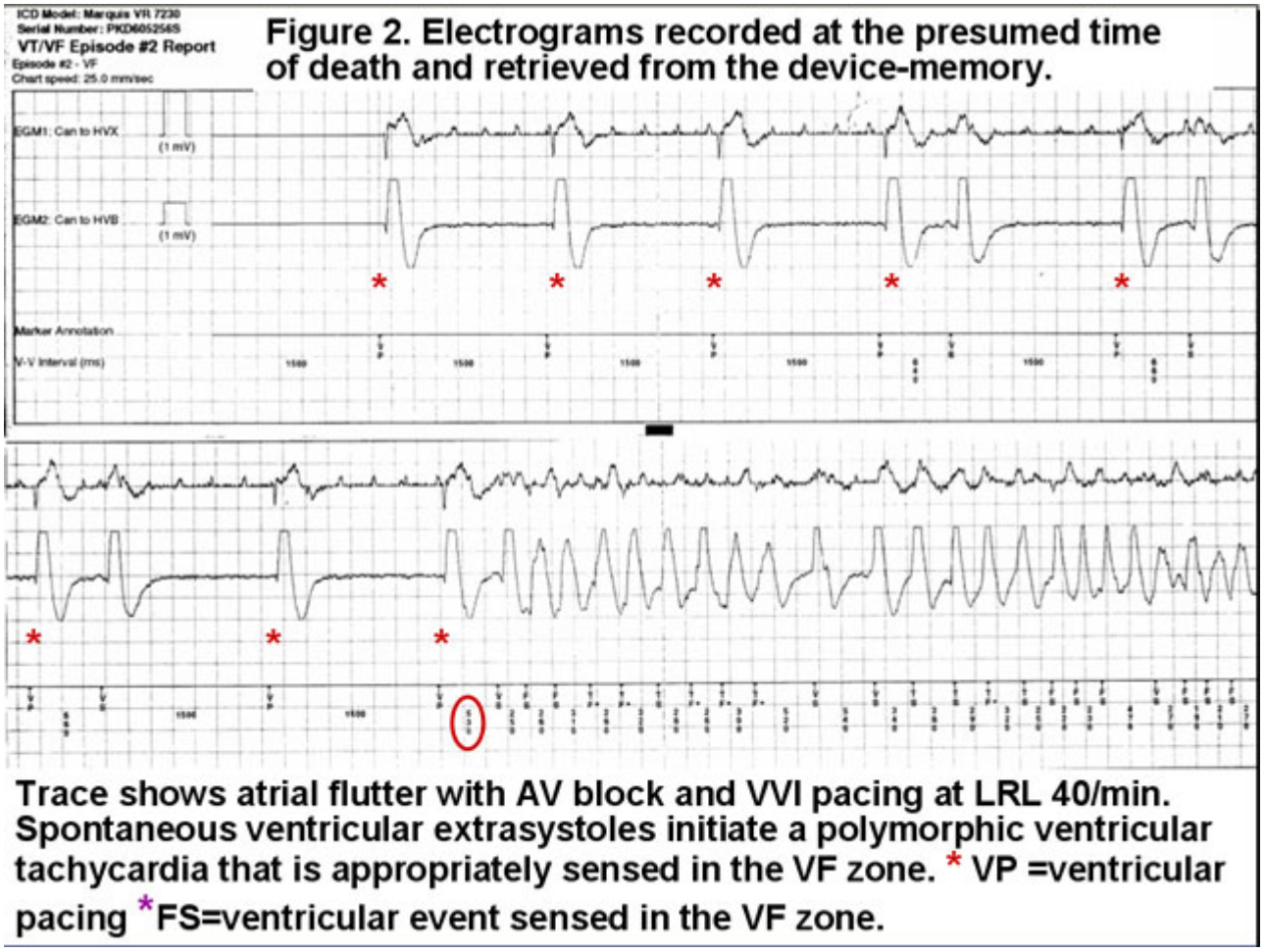
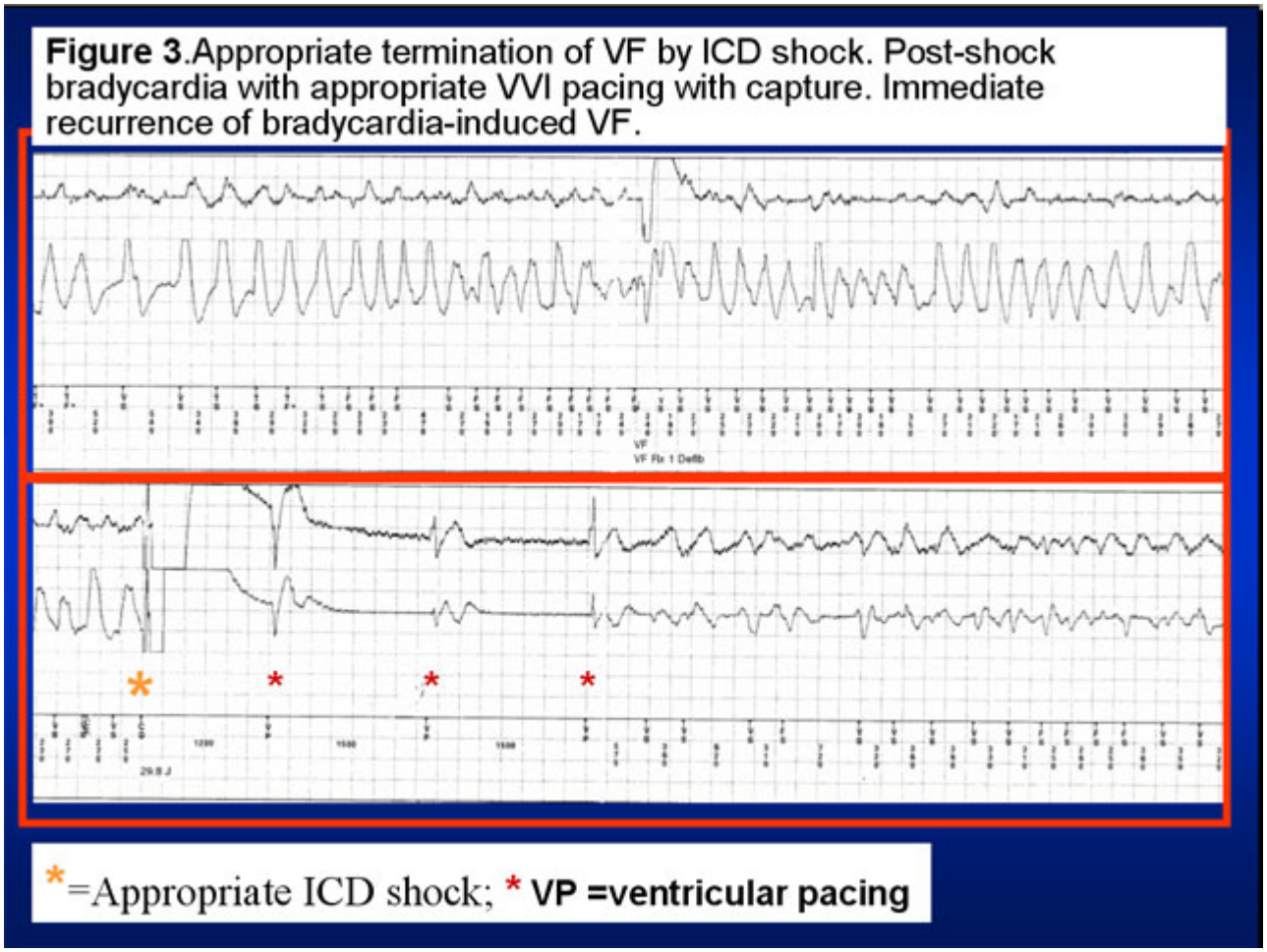


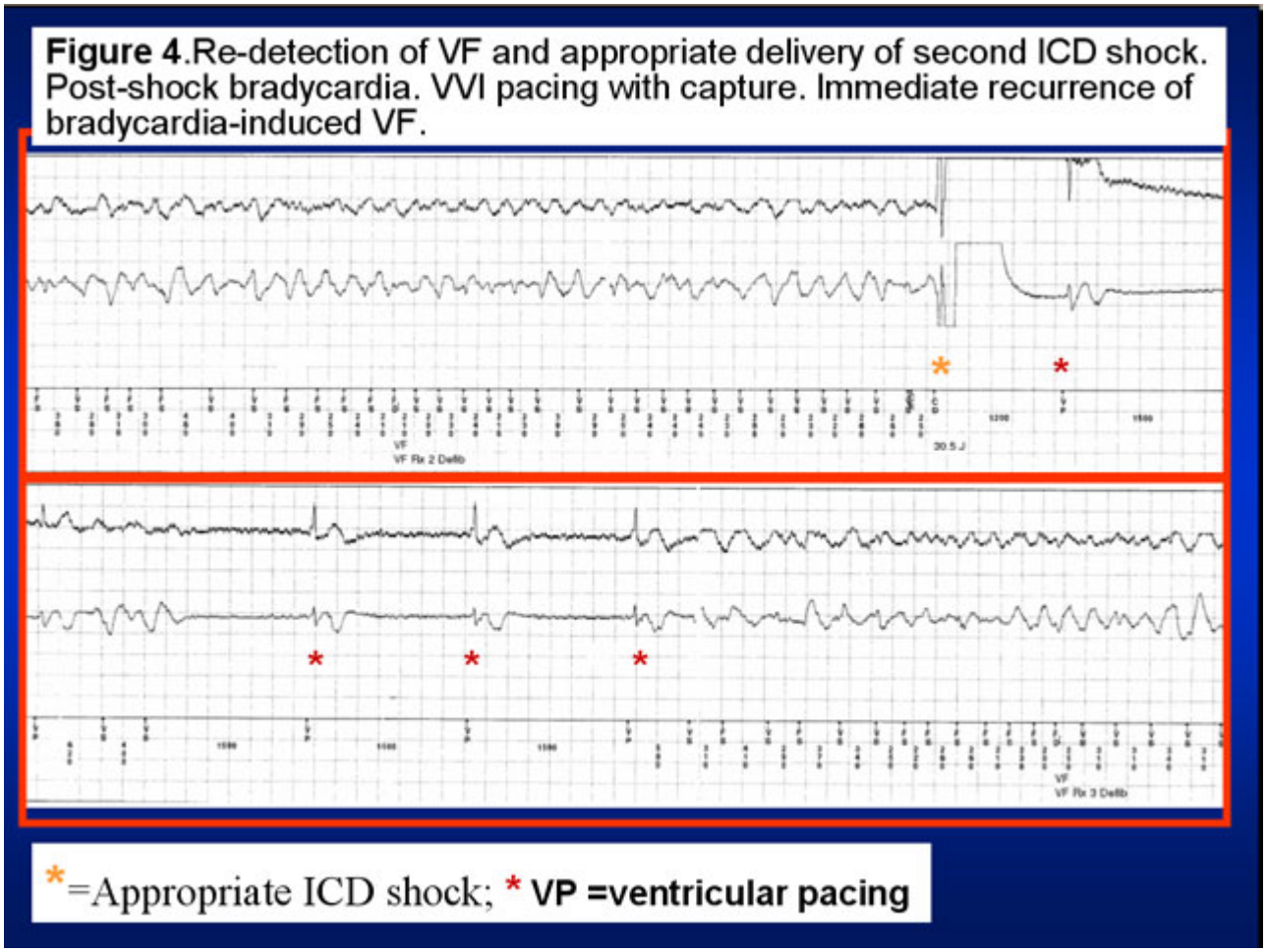
Figure 2. Electrograms recorded at the presumed time of death and retrieved from the device-memory.

Trace shows atrial flutter with AV block and VVI pacing at LRL 40/min. Spontaneous ventricular extrasystoles initiate a polymorphic ventricular tachycardia that is appropriately sensed in the VF zone. * VP =ventricular pacing *FS=ventricular event sensed in the VF zone.

The device delivers an appropriate ICD shock that successfully terminates the tachycardia (figure 3). The shock also converts the atrial flutter to atrial fibrillation, yet AV block persists and ventricular pacing at 40 beats/min is delivered after the shock (figure 3).



Following the third paced complex, polymorphic VT starts again. In figure 4 a similar sequence of events is recorded: VF terminated by ICD shock, ventricular pacing at the lower rate limit and immediate re-initiation of polymorphic VT.



In figure 5 the traces are not continuous. The figure shows two additional ICD shocks. Both shocks are appropriate and successful in terminating VF. Figure 5 also shows that the QRS complexes widen during pacing and during VF. One episode of polymorphic VT actually terminates spontaneously (lower panel).

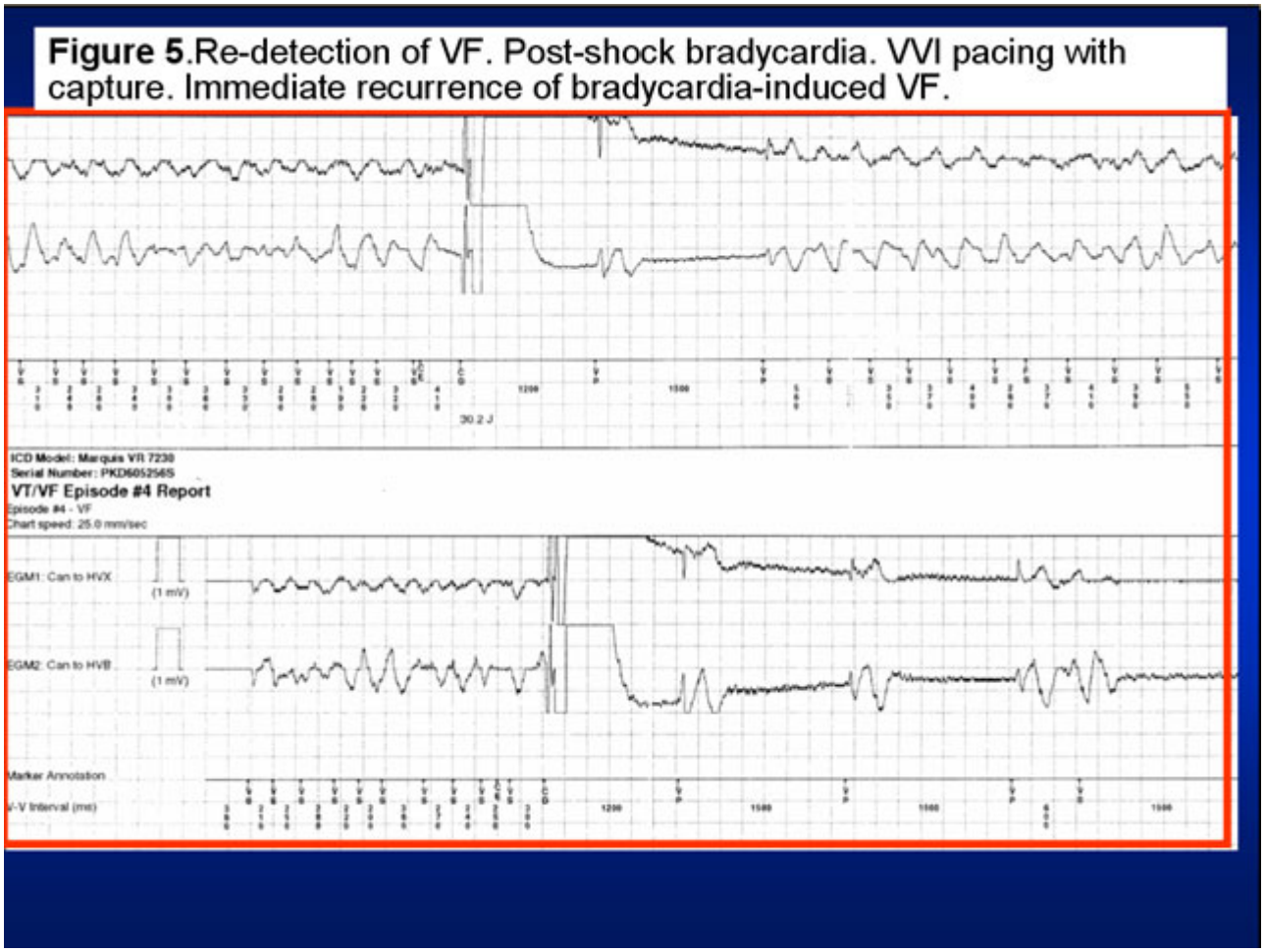


Figure 5.Re-detection of VF. Post-shock bradycardia. VVI pacing with capture. Immediate recurrence of bradycardia-induced VF.

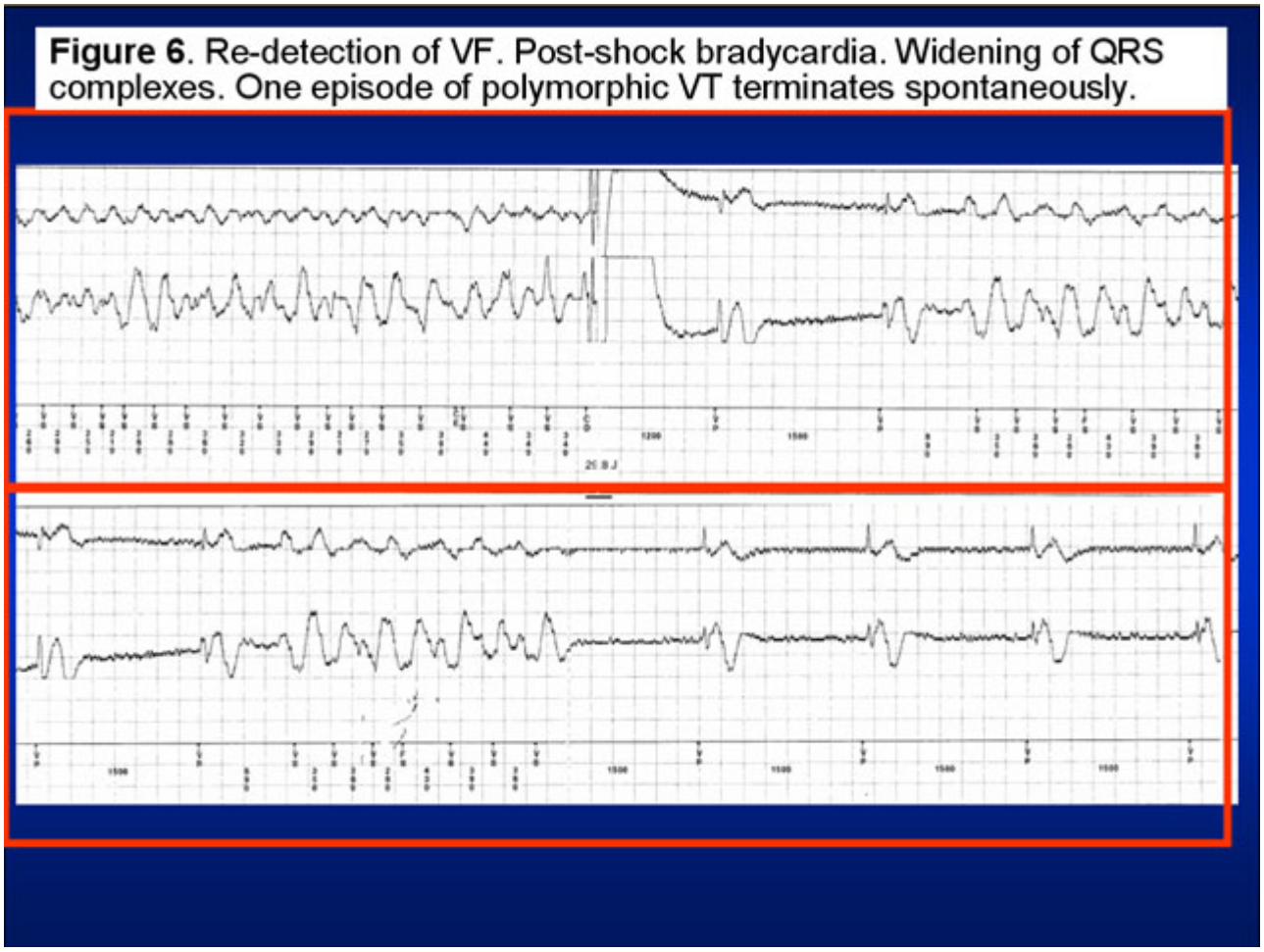


Figure 6. Re-detection of VF. Post-shock bradycardia. Widening of QRS complexes. One episode of polymorphic VT terminates spontaneously.

However, the terminal arrhythmia develops, as depicted in figure 7: VF is not detected by the device, presumably due to the declining amplitude of the local intracardiac electrograms after numerous intracardiac shocks. The patient died in VF.

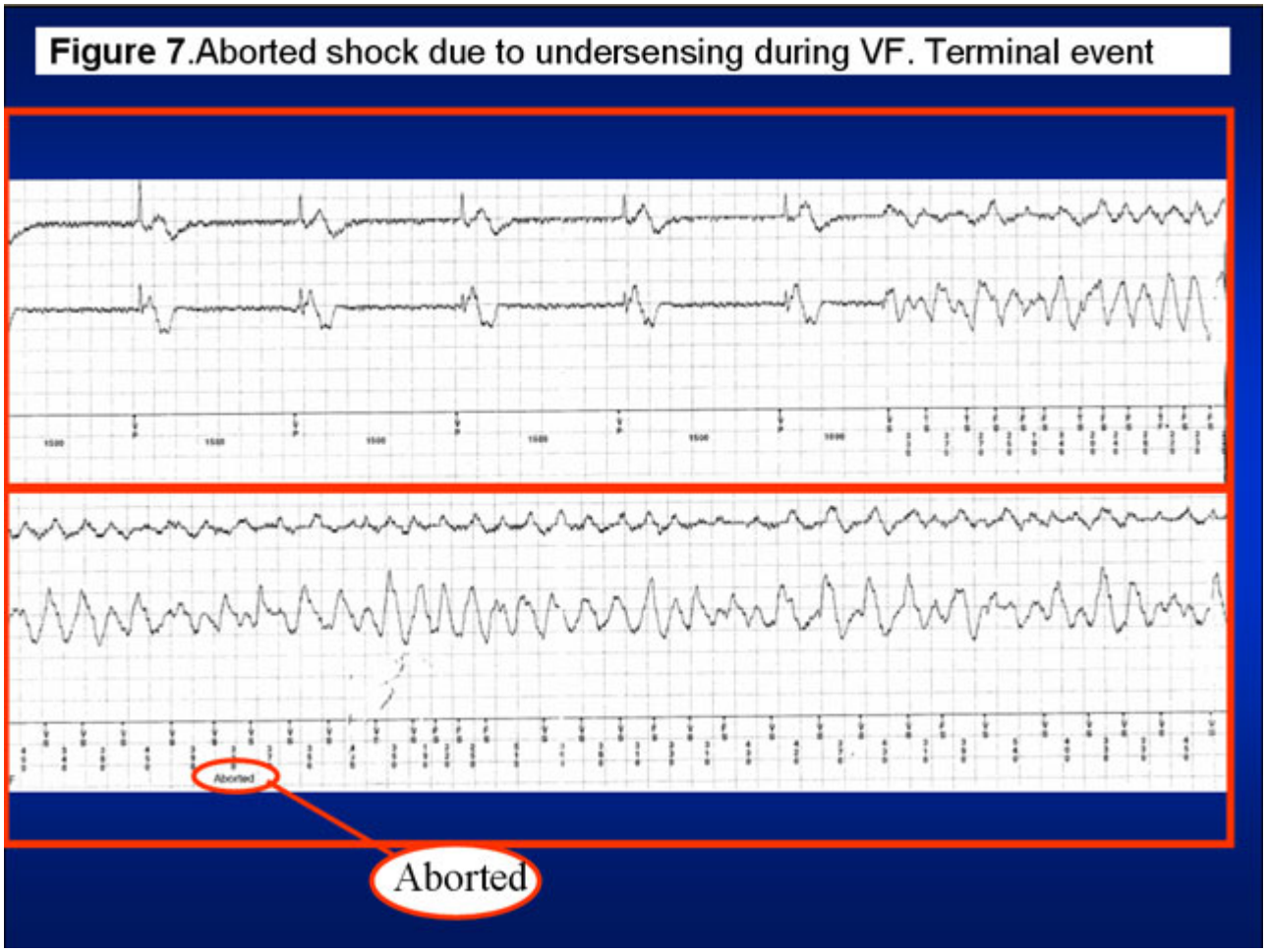


Figure 7.Aborted shock due to undersensing during VF. Terminal event

Discussion. Although the QT interval cannot be accurately measured or even appreciated from the retrieved intracardiac electrograms, the fact that this is a polymorphic VT occurring in the setting of severe bradycardia strongly suggests the diagnosis of pause-dependent (bradycardia-dependent) torsade de pointes. Further supportive evidence for this diagnosis is derived from the device annotations shown in figure 2. Note that the coupling interval of the first tachycardia interval (the interval between the last paced complex and the first VT-complex) is 530 msec (red circle in figure 2). This is a coupling interval in the range seen in the long QT syndrome (1).

The AV block that precipitated the torsade was unexpected. We were later able to find only anecdotal reports of AV-block in patients with HIV-related cardiomyopathy treated with anti-retroviral therapy (2,3). The HIV-protease inhibitors probably contributed to the onset of torsade de pointes during AV block because these drugs block the IKr potassium-channels and are therefore a recognized cause of long QT syndrome (4).

The torsade de pointes was appropriately detected and treated by the ICD. However, the bradycardia persisted and therefore, an arrhythmic storm of pause-dependent torsade de pointes ensued. This arrhythmic storm could have been prevented by more rapid pacing. ICDs have a “post-shock pacing” function, that can be programmed differently from the basic pacing features. In all ICDs “post-shock pacing” is delivered at higher voltage to ensure ventricular capture in the face of the temporary rise in local pacing threshold that invariably follows an intracardiac shock. Additionally, many devices have a “post-shock pacing” mode than can be programmed to pace faster than the basic pacing rate. For patients considered to be at risk for torsade de pointes, either due to a baseline long QT or because they receive QT prolonging mediations, we usually program the devices to pace at 100-120 beats/min for 10 – 60 minutes after each shock to prevent the sequence of events seen here. Unfortunately, this particular device (and all but the newest Medtronic ICDs) does not have an independently programmable post-shock pacing rate. Therefore, pacing continued at high voltage but at a fixed rate after each shock and pause-dependent torsade recurred immediately after each shock.

The tragic ending of this case taught us several lesions: 1) Use of HIV protease inhibitors entails a risk for QT prolongation (4), a side effect we were unaware during the patient’s hospitalization. 2) We certainly did not expect that AV block would occur and trigger torsade de pointes. Had we performed an EP-study before ICD implantation it is possible (albeit far from certain) that a prolonged H-V interval would have been recorded. In that case, a device with faster post-shock pacing could have been selected. 3) At the time, we had ardent deliberations regarding the need for ICD implantation in this particular patient because of the relatively preserved left ventricular function and the comorbidities discussed in Part II. We never received any protests (by any of the

physicians involved) following the tragic ending of this case. Apparently, it is acceptable to die suddenly as long as one has an implanted ICD. My guess is that this part of the story (the lack of accusations) would have been different if the patient had not been treated “according to the guidelines.”

References for Part III.

1. Viskin S, Alla SR, Barron HV, et al. Mode of onset of torsade de pointes in congenital long QT syndrome. J Am Coll Cardiol 1996;28:1262-8.
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3. Lipshultz SE, Chanock S, Sanders SP, Colan SD, Perez-Atayde A, McIntosh K. Cardiovascular manifestations of human immunodeficiency virus infection in infants and children. Am J Cardiol 1989;63:1489-97.
4. Anson BD, Weaver JG, Ackerman MJ, et al. Blockade of HERG channels by HIV protease inhibitors. Lancet 2005;365:682-6.