A 36-year-old woman was referred after her son had syncopal episodes associated with a markedly prolonged QT interval, which was treated with β-blockers and a subsequent pacemaker. The mother had a history of “seizures” as a child that had been treated with various medications, which did not appear to affect her events. She continued to have episodes 2 to 3 times a year, and witnesses said she would “stiffen up and gasp for air.” She had had no syncope for many years but had episodes of dizziness and near syncope when anxious or under emotional stress. These episodes were not associated with palpitations. Her ECG revealed a corrected QT interval of 496 (Bazzett’s formula), and an echocardiogram was remarkable for moderate mitral regurgitation without prolapse. Her QT interval did not shorten during treadmill testing. During recovery, she demonstrated biphasic inferolateral T-wave changes that were more pronounced with hyperventilation. On the basis of her long QT interval, she was placed on a β-blocker, and in view of her near syncopal symptoms, an event recorder was placed.

Most of her strips were unremarkable except for an occasional premature ventricular contraction. On one occasion when she became anxious and hyperventilated in a parking lot, she recorded the strip seen in Figure 1, showing marked T-wave alternans. On the basis of data from the International Long QT Registry,1,2 implantation of an implantable cardioverter-defibrillator was recommended. This was done, and 8 months later, while in a stressful situation (and on β-blockers), the patient had the episode of polymorphic ventricular tachycardia shown in Figure 2, which was associated with syncope and required multiple shocks from her device.

References

Figure 1. Progressive T-wave alternans from event recorder.
Figure 2. Intracardiac electrogram from patient's implantable defibrillator showing onset of polymorphic ventricular tachycardia followed by pretherapy tracing (confirmation) before first defibrillation.
Malignant T-Wave Alternans
Daniel S. Goldman, Wojciech Zareba and Arthur J. Moss

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