Twiddler syndrome refers to a rare complication observed in patients with implanted pacemakers or defibrillators that causes device malfunction. The condition arises as a result of the patient’s conscious or unconscious manipulation of the device, causing its rotation in the pocket, resulting in torsion and dislodgment of the implanted lead(s).

A 65-year-old obese woman with idiopathic dilated cardiomyopathy and diabetes mellitus had undergone implantation of a biventricular implantable cardioverter-defibrillator (CRT-D, Medtronic Insync Maximo 7304) with the positioning of 3 leads: a passive bipolar lead in the auricle of the right atrium (Medtronic Capsure Z Novus 5554), a bipolar, single-coil, screw-in lead (Medtronic Sprint 6931) in the apex of the right ventricle (RV), and a unipolar left-ventricular (LV) pacing lead (Medtronic Attain 4193) in an anterolateral tributary of the coronary sinus.

Approximately 1 month after implantation, the patient presented to the emergency room with effort dyspnea followed by gradually worsening hiccups associated with dysphonia (a metallic or “pseudo-robot” voice) was observed. The device was investigated: During threshold testing of the RV, no effective capture could be observed, even at maximum output. In contrast, when testing the unipolar LV lead (which in this model closes off on the RV coil), the symptoms of hiccups and “pseudo-robot” voice were immediately reproduced. The device was reprogrammed in VVI mode 30 bpm so that underlying spontaneous sinus rhythm emerged, and symptoms ceased immediately.

Chest radiography (Figure 1) showed a macrodislodgment of the screw-in RV coil lead into the brachiocephalic vein (at the level of the esophagus) and an impressive cluster of catheter tangles in the subcutaneous pocket. The right atrium lead, although slightly retracted and with a large loop, appeared to have the tip in the correct position, with good sensing and pacing values. The LV unipolar lead (with the dipole closing on the RV coil in the brachiocephalic vein) initially presented a slight retraction but with good sensing and pacing threshold.

Four days later, the patient underwent a revision procedure aiming at repositioning both the LV and RV coil leads. The chest radiograph at that point (Figure 2) revealed further retraction of both the RV and LV leads (no LV pacing was possible at that point). After incision into the skin and gentle extraction of the device, an impressive cluster of twisted catheter tangles connected to the device (Figure 3) was found. The device had rotated innumerable times on its central axis. As a consequence of the numerous loops and twists, both the RV lead coil and the LV lead had been deformed and needed replacement.

Twiddler syndrome is a rare and potentially dangerous cause of lead dislodgment in patients with pacemakers and implantable cardioverter-defibrillators. The peculiar clinical presentations of Twiddler syndrome in conventional pacemakers include abdominal pulsation and stimulation of the pectoral major muscle. Patients who are most at risk for this condition include middle-aged obese women and patients with mental disorders such as dementia. To our knowledge, this is the first report of Twiddler syndrome in a CRT-D device with retraction of a screw-in defibrillation lead. The unique symptoms of hiccups and dysphonic metallic voice have never been reported before and were the result of the particular technical characteristics of the Medtronic CRT-D device system, in which the unipolar tip of the LV lead uses the RV defibrillator coil as the anode. The macrodislodgment of the RV coil (retracted into the brachiocephalic vein, at the level of esophagus) with consequent anatomic proximity to both recurrent and phrenic nerves caused an electrical irritation of these nerves, hence explaining the very odd symptoms. Treatment measures to avoid or limit Twiddler syndrome in patients at increased risk include limiting the pocket size and the suturing of the device to the fascia.

Disclosures

None.

References

Figure 1. Chest radiograph shows the configuration of electrodes when the patient presented in the emergency room with hiccups and a dysphonic metallic voice. ATR indicates atrial bipolar lead; RV, screw-in right ventricular coil lead; and LV, left ventricular electrode in the coronary sinus.

Figure 2. Chest radiography performed a few days later showed further retraction of both the RV and LV leads. ATR indicates atrial bipolar lead; RV, screw-in right ventricular coil lead; and LV, left ventricular electrode in the coronary sinus.

Figure 3. Intraoperative image showing the cluster of catheter tangles found in the device pocket.
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Circulation. 2006;114:e534-e535
doi: 10.1161/CIRCULATIONAHA.106.626788

Circulation is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
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Print ISSN: 0009-7322. Online ISSN: 1524-4539

The online version of this article, along with updated information and services, is located on the World Wide Web at:
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