



## Review

### Twiddler's Syndrome: 50 years and still counting (or twirling)!

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It is said that human beings are creatures of habits. As such human behavioral patterns and acts are controlled either by volition or involuntary processes. Habits can also cast direct or indirect influence on a plethora of human ailments and suffering. This article describes observations made on yet another malady caused by human habits and repetitive behavioral patterns— namely Twiddler's syndrome.

#### Patient 1

A 46-year old man underwent implantation of a biventricular pacemaker – defibrillator in the left pre-pectoral region for severe cardiomyopathy, congestive heart failure and high degree atrioventricular block. Perioperative course was unremarkable. Six weeks post implant, the patient presented with wound dehiscence and pocket infection. As conservative management of the device pocket with wound care and antibiotic therapy did not result in eradication of the infection, the patient underwent removal of the entire device system. Surgical opening of the pocket revealed evidence of repetitive manipulation of the device system (figure 1). The pulse generator was found immediately below the incision, as opposed to its original placement deep in the subcutaneous tissue, above the pectoralis major muscle. All implanted leads were curled together. Anchoring plastic sleeves and sutures were dislodged from the underlying pectoralis fascia and muscle. Leads were about to be dislodged from their myocardial attachment sites – denoting an impending disaster in a patient who is dependent on pacing. A new biventricular pacemaker – defibrillator system was implanted in the right pre-pectoral region. The patient vehemently denied voluntary manipulation of the device pocket.

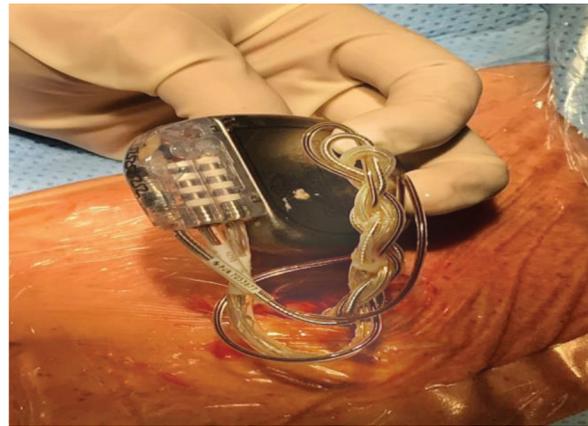
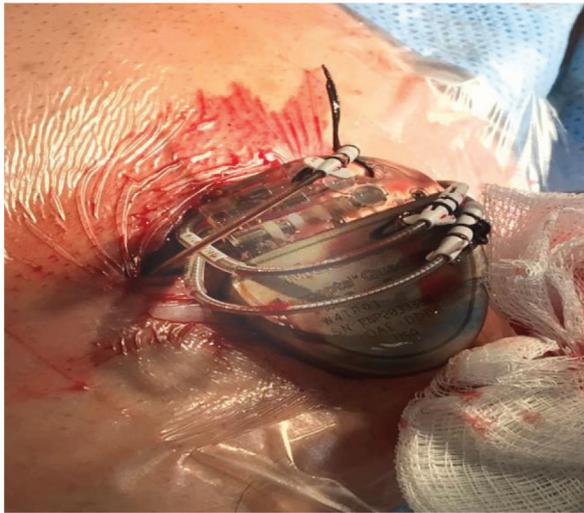


Figure 1

#### Patient 2

A 68 year old woman underwent implantation of a biventricular pacemaker system for severe non-ischemic cardiomyopathy and congestive heart failure. Intra operatively, the patient had satisfactory pacing and sensing parameters in all leads. At 8 weeks post implant, the patient presented with persistent jolting sensation in the left upper quadrant of the abdomen due to inadvertent left phrenic nerve capture. A chest x-ray demonstrated dislodgement of the left ventricular pacemaker lead and straightening of right atrial and right ventricular leads. Surgical opening of the pocket revealed coiling of the leads around the pulse generator, resulting in lead dislodgment (figure 2). Even in this case, the patient did not acknowledge deliberate manipulation of the device.



**Figure 2**

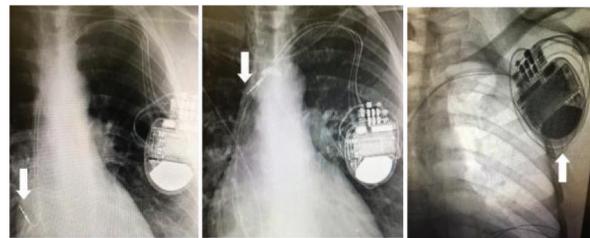
### Patient 3

A 75 year old man underwent implantation of a dual chamber pacemaker for sick sinus syndrome. Post operatively, he developed erythema around the skin incision in the left chest. A superficial pocket infection was suspected. Oral antibiotic therapy for one week resulted in only temporary suppression of erythema. A chest x-ray suggested migration of the pacemaker pulse generator caudally. Both the atrial and ventricular leads no longer exhibited satisfactory slack and redundancy that were apparent at the time of the procedure. The patient's daughter acknowledged that her father was involuntarily manipulating the pacemaker pocket on certain occasions such as watching television. After the clinic visit, she bought her father a necklace and advised him to twist it, instead of twirling the pacemaker generator. After introduction of the substitute "play item", the pocket infection resolved within 2 weeks.

### Patient 4

A 46 year old woman underwent implantation of a biventricular pacemaker-defibrillator for severe non-ischemic cardiomyopathy and congestive heart failure despite optimal medical therapy. The patient had an elaborate skin tattoo carved on the chest. At the time of device implantation, every effort was made to avoid placement of the incision over the tattoo. Two weeks after the procedure, the patient presented with intermittent twitching in the upper left pectoral region.

There was no evidence for pocket infection or wound dehiscence. Device interrogation revealed absence of atrial sensing. A chest radiograph showed retraction of the atrial pacemaker lead and increased coiling of leads around the pulse generator (figure 3). Two days later, the patient was taken back to the electrophysiology laboratory for atrial lead revision. Prior to re-opening of the pocket, fluoroscopic examination showed complete retraction of the atrial lead in to the pocket. Interestingly, both right and left ventricular leads remained secured and this was attributed to the redundant lengths of these two leads in the pocket. The patient reluctantly admitted a prior habit of twiddling over the tattoo.



**Figure 3**

(Left) Chest x-ray 6 hours after implantation of the biventricular pacemaker – defibrillator showing atrial lead appropriately anchored to the lateral right atrium. (Middle) Chest x-ray 14 days weeks after device implantation revealing retraction of the atrial lead in to the left innominate vein – superior vena cava junction. Also note increased coiling of redundant leads around the generator. (Right) Fluoroscopy 16 days after device implantation, showing complete absence of the atrial lead in the intra vascular space and lead coiling behind the generator.

### Discussion

Oxford living dictionary defines Twiddler as a person who twists, moves or fiddles an object, typically in a purposeless or nervous way<sup>(1)</sup>. Twiddler's syndrome refers to twisting and twirling of an implantable device such as a pacemaker or defibrillator pulse generator by the "host", resulting in a complication or device malfunction<sup>(2)</sup>. Since its first description fifty years ago in 1968, various permutations of Twiddler's syndrome have been described in the medical literature<sup>(3)</sup>.

Twiddler's syndrome is generally not considered a common clinical entity. It is usually associated with painless lead dislodgement and often diagnosed within the first year of implant.



Complications of Twiddler's syndrome are due to logical consequence of spinning of the pulse generator in the pocket and the results can be potentially lethal. These complications include pocket infection, lead dislodgment, cessation of pacing, delivery of inappropriate defibrillator shocks, diaphragmatic twitching due to stimulation of the phrenic nerve or hemidiaphragm, pleuritic or pericarditic chest pain, vascular perforation and accidental extraction of the device system. With continuous wrapping of the leads around the generator, muscle twitching in the arm could occur, due to electrical stimulation of brachial plexus.

Some authors have noted that elderly and obese patients are at an increased risk because of loose subcutaneous tissue, which allows rotation of the pulse generator in the pocket<sup>(2)</sup>. Our observations suggest that patients with dementia, prior psychiatric illness, chemical dependency and those who live alone without a life partner are also prone to manipulate the devices deliberately or involuntarily.

Beside clues provided by patient's history and physical examinations, chest radiograph may provide useful information such as rotation or migration of the pulse generator and retraction of the leads.

Management of Twiddler's syndrome requires multi-pronged approach that includes patient counseling and frequent follow up, referral to psychiatry when indicated, employment of substitutes for twiddling such as necklaces. Modified surgical techniques such as creation of a smaller surgical pocket, use of active fixation leads, suturing the pulse generator to the pectoralis fascia and implantation of the device in the sub-pectoral region may discourage or prevent manipulation of the pulse generator and the leads<sup>(3)</sup>.

In our experience, use of antimicrobial envelopes around the pulse generator could potentially reduce the risk of infections associated with twiddling. Recent introduction of leadless pacemakers could also serve as an option for those patients who develop refractory symptoms despite adoption of the above mentioned measures.

Twiddler's syndrome is not confined to cardiac implantable electronic devices. It has been described in situations of non-cardiac medical devices such as spinal cord stimulators and chemotherapy infusion pumps<sup>(4)</sup>.

Due to widespread implantation of medical devices for various indications, the number of cases of Twiddler's syndrome is likely to increase exponentially. As a result, contemporary clinicians are likely to benefit from maintaining a high degree of suspicion for Twiddler's syndrome in appropriate circumstances.

## References

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