

What's the Twist? Twiddler's Syndrome in Deep Brain Stimulation

Zane Tymchak, Aleksander Vitali

Keywords: Brain stimulation, Movement disorder surgery

doi:10.1017/cjn.2017.230

Can J Neurol Sci. 2017; 44: 726-727

Our patient is a 61-year-old female who underwent repeat bilateral ventrointermediate nucleus implantation of a deep brain stimulation (DBS) system for essential tremor. Previously, she had been implanted with a DBS system, but this was explanted secondary to infection. This second surgery was successful and resulted in a marked reduction of her tremors. She presented 3 months postoperatively with complaints of pain and tenderness in the left side of her neck. Plain radiographs revealed twisting of the extension leads in the neck (Figure 1A,B). She was taken for successful revision of these extension leads; however, she presented again 3 weeks later with purulent drainage from her subclavicular pouch and tenderness along the extension lead tract. She was taken back to surgery, where the entire system was found to be contaminated and was therefore explanted. We have decided to not reimplant the system at this time.

Twiddler's syndrome in the neuromodulation population is a rare but important cause of hardware failure, occurring in approximately 1% of cases.^{1,2} This phenomenon was originally reported in 1968 in the setting of implantable cardiac devices.³ Since then, it has become a recognized complication of implantable devices, including both DBS and spinal cord stimulation (SCS) systems.^{1,2,4} As its name suggests, it is thought to be due to direct patient manipulation of the pulse generator, although this has been questioned by some.⁵ Known risk factors include psychiatric illness, obesity, and age.^{5,6} Patients typically present

with early or late reemergence of symptoms, which can be accompanied by wound complications, localized pain, and tenderness of the lead tract, as well as elevated impedance recordings on routine follow-up.^{1,2,5-7} Diagnosis is usually made with plain radiographs. Imaging of the brain should be considered, as severe cases can result in intracranial electrode retraction.^{5,8,9} Treatment options include revision surgery or explantation of the neuromodulation system, though this is usually only reserved for cases of secondary infection, as was the case in our patient. Revision surgery can involve simple resuturing of the pulse generator within its subcutaneous pouch; however, some cases may require more extensive revision and replacement of hardware if damage has been incurred to the leads or the intracranial electrode has been withdrawn.

DISCLOSURES

The authors hereby state that they have no conflicts of interest to disclose.

REFERENCES

1. Burdick AP, Okun MS, Haq IU, et al. Prevalence of Twiddler's syndrome as a cause of deep brain stimulation hardware failure. *Stereotact Funct Neurosurg.* 2010;88(6):353-9.
2. Al-Mahfoudh R, Chan Y, Chong HP, Farah JO. Twiddler's syndrome in spinal cord stimulation. *Acta Neurochir (Wien).* 2016;158(1):147-54.
3. Bayliss CE, Beanlands DS, Baird RJ. The pacemaker-Twiddler's syndrome: a new complication of implantable transvenous pacemakers. *Can Med Assoc J.* 1968;99(8):371-3.
4. Morishita T, Foote KD, Burdick AP, et al. Identification and management of deep brain stimulation intra- and postoperative urgencies and emergencies. *Parkinsonism Relat Disord.* 2010;16(3):153-62.
5. Silva PA, Chamadoira C, Costa H, Linhares P, Rosas MJ, Vaz R. Twiddler (or not) syndrome: questioning etiology for an uncommon form of hardware malfunction in deep brain stimulation. *Surg Neurol Int.* 2014;5(Suppl 8):S410-2.
6. Astradsson A, Schweder PM, Joint C, Green AL, Aziz TZ. Twiddler's syndrome in a patient with a deep brain stimulation device for generalized dystonia. *J Clin Neurosci.* 2011;18(7):970-2.

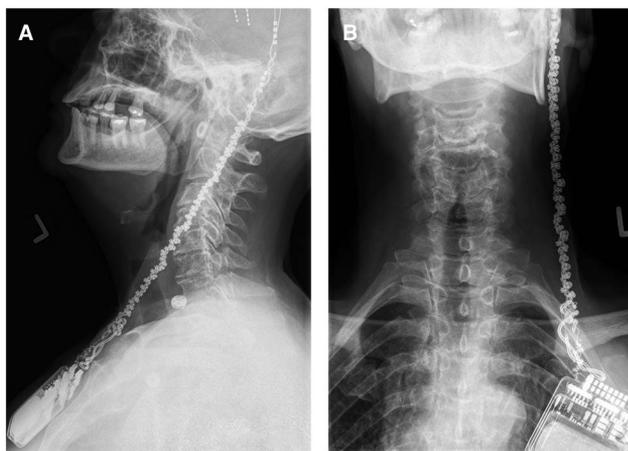


Figure 1: Lateral (A) and AP (B) x-rays showing extensive twisting of DBS extension leads.

From the Department of Surgery, Division of Neurosurgery, University of Saskatchewan, Saskatoon, Saskatchewan, Canada (ZT, AV).

RECEIVED MARCH 21, 2017. FINAL REVISIONS SUBMITTED JUNE 5, 2017. DATE OF ACCEPTANCE JUNE 11, 2017.

Correspondence to: Aleksander Vitali, Department of Surgery, Division of Neurosurgery, University of Saskatchewan, Royal University Hospital, 103 Hospital Drive, Saskatoon, Saskatchewan, Canada S7N 0W8. Email: alex.vitali@usask.ca.

7. Moens M, Petit F, Goudman L, et al. Twiddler's syndrome and neuromodulation devices: a troubled marriage. *Neuromodulation*. 2016;20(3):279-83.
8. Gelabert-Gonzalez M, Relova-Quinteiro JL, Castro-Garcia A. "Twiddler syndrome" in two patients with deep brain stimulation. *Acta Neurochir (Wien)*. 2010;152(3):489-91.
9. Sobstyl M, Ząbek M, Górecki W, Brzuszkiewicz-Kuźmicka G. Twiddler syndrome in a patient with tremor-dominant Parkinson's disease: a case report and literature review. *Neurol Neurochir Pol*. 2015;49(6):467-71.