

A Rare Hardware-related Complication After Deep Brain Stimulation: Two Cases of Twiddler's Syndrome

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Abstract

Twiddler's syndrome is an uncommon complication of deep brain stimulation, arising from the inadvertent rotation of the implanted pulse generator, which results in lead coiling, displacement, or fracture. This results in stimulation failure and symptom recurrence, often necessitating surgical revision. While well-documented in cardiac devices, Twiddler's syndrome remains infrequent in operations. Early diagnosis through radiographic imaging and impedance abnormalities is crucial for prompt intervention.

We report two cases of Twiddler's syndrome in patients with movement disorders: a 73-year-old male with Parkinson's disease and a 55-year-old female with multifocal dystonia, both of whom had bilateral deep brain stimulation with left-sided dual-channel implanted pulse generator. After years of effective symptom control, both patients presented with worsening motor symptoms. Device examination revealed abnormal impedance readings, and imaging confirmed extensive lead coiling. Surgical revision was performed, including lead replacement and implanted pulse generator repositioning under the pectoralis fascia. Both patients demonstrated significant symptom improvement postoperatively, with restored impedance levels and functional recovery.

Twiddler's syndrome is a rare but critical complication of deep brain stimulation, leading to hardware failure and clinical deterioration. Early diagnosis through imaging and impedance monitoring is essential. Preventive strategies, including secure implanted pulse generator fixation and careful pocket sizing, are key to minimizing risk and improving patient outcomes.

Keywords: deep brain stimulation, movement disorders, Twiddler's syndrome, hardware complications, extension fracture.

Introduction

Twiddler's syndrome is a rare but serious hardware-related complication following deep brain stimulation (DBS) surgery. It is characterized by unintended rotation of the implanted pulse generator (IPG) within its subcutaneous pocket, leading to excessive coiling, displacement, or fracture of the leads [1]. This results in stimulation failure, loss of therapeutic benefit, or even neurological worsening.

Originally described in cardiac pacemakers by Bayliss in 1968, Twiddler's syndrome has since been reported in various neurostimulation devices, including DBS [2].

Although well-documented in cardiac implantable electronic devices, where it occurs in approximately 1.2% of cases [3], Twiddler's syndrome remains an infrequent

but clinically significant complication in DBS, with an estimated prevalence of 0.07% to 1.4%. [4–6].

The condition is often associated with rapid symptom recurrence, stimulation inefficacy, and abnormal impedance readings. Radiographic evaluation, particularly chest X-ray, may reveal coiled, displaced, or fractured leads—often in a double-helical pattern [7].

Several risk factors have been implicated, including an oversized implantation pocket, insufficient device fixation, and patient-related behaviors such as compulsive manipulation of the IPG. Additional predisposing factors identified in cardiac patients—such as female sex, high body mass index (BMI), and antidepressant use—may also be relevant in the DBS population [3, 5].

From a neurosurgical perspective, Twiddler's syndrome presents both diagnostic and management

challenges. Lead displacement compromises clinical outcomes and often necessitates surgical revision. Prevention strategies, including optimal IPG placement, secure anchoring techniques, and patient education on device care, are critical in minimizing this complication. Despite these considerations, Twiddler's syndrome in DBS remains underreported, limiting consensus on best management practices.

While direct evidence linking individual genetic and neurobiological factors to hardware-related complications in neuromodulation therapies is currently limited, emerging research suggests that patient-specific factors, including genetic predispositions and neurobiological responses, may influence outcomes. For instance, genetic factors influencing BMI and compulsive behaviors have been implicated in conditions like obsessive-compulsive disorder (OCD), which shares phenotypic similarities with behaviors that could lead to hardware manipulation. Furthermore, variations in genes related to neurotransmitter systems, particularly those involved in serotonin and dopamine signaling, have been associated with compulsive behaviors. Some of the key genetic factors include: **HTR2A** and **HTR1B** (serotonin signaling), as well as **DRD2** and **DAT1** (dopamine regulation). These genetic insights suggest a potential link between neurobiological predispositions and hardware-related complications in neuromodulation therapies. However, further investigation is warranted to clarify their specific roles and clinical implications in patient management [8].

In this report, we present two cases of Twiddler's syndrome in movement disorder patients: one with Parkinson's disease undergoing bilateral subthalamic nucleus (STN) DBS and another with multifocal dystonia undergoing bilateral globus pallidus internus (GPi) DBS. Through these cases, we aim to highlight the clinical course, radiographic findings, surgical management, and preventive strategies for this uncommon complication. Over an 11-year follow-up period, during which 620 neuromodulation procedures were performed, these cases remained exceptionally rare.

Case presentation

Illustrative case 1. A 73-year-old male patient with a Stage 5 Parkinson's Disease diagnosis according to the Hoehn & Yahr scale underwent bilateral STN DBS using a left dual-channel IPG (Activa PC). The initial programming of the DBS system resulted in significant improvement in symptoms for over 6 years.

Recently, the patient experienced discomfort in the left clavicular region and a gradual worsening of his condition, including increased stiffness and tremors in the limbs and a decrease in self-care skills over a three-week period. Upon evaluation, impedance and current measurements for electrodes 0, 1, and 2 were within normal ranges with current output (4.1mA), suggesting that the circuitry was intact, however, electrode 3 displayed low impedance level (234), indicating a potential problem with the lead.

To determine the cause of the suspected malfunction, the patient underwent diagnostic imaging, including cranial and chest X-rays. The X-rays revealed twisting of the IPG and extension leads around their axis (figure 1a-c).

A surgical intervention was performed, during which twisting of the DBS extension leads was observed, necessitating replacement of the extension leads. Additionally, the IPG was repositioned under the pectoralis fascia to reduce the risk of future displacement and coiling.

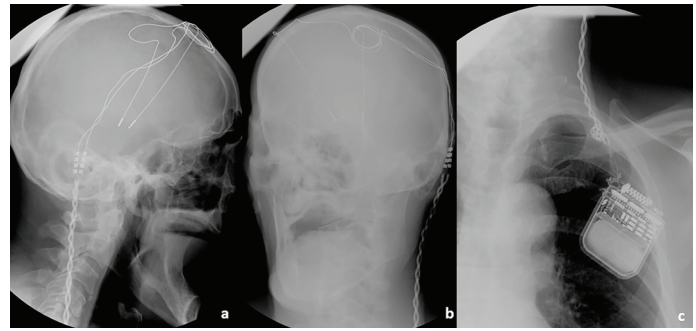


Figure 1 – Cranial (a, b) and Chest (c) X-ray show coiling of extension wires.

This intervention significantly improved the patient's symptoms, and follow-up examinations showed that impedance levels had returned to normal.

Illustrative case 2. The patient is a 55-year-old female diagnosed with multifocal dystonia, which has manifested as cervical and torsion dystonia for the past 11 years. She has experienced significant improvement in symptoms following bilateral GPi DBS and the placement of a left dual-channel IPG (Activa PC). This treatment has effectively managed her condition for ten years. The IPG has required battery replacements 5.5 years and 3.5 years prior due to low capacity of battery.

The patient was readmitted to the hospital due to worsened uncontrolled muscle cramps, spasms, and difficulties with speaking and swallowing, indicating a worsening of her dystonia.

During evaluation, impedance and current measurements for electrodes 0, 1, and 2 were within normal ranges, suggesting the circuitry was intact. However, electrode 3 showed significantly increased impedance levels (10700-12400) and decreased current output (0.6mA), indicating a potential issue with the lead. Subsequent chest X-rays (figure 2a) revealed extensive coiling of the DBS extension leads, a mechanical complication that could affect the effectiveness of the DBS.

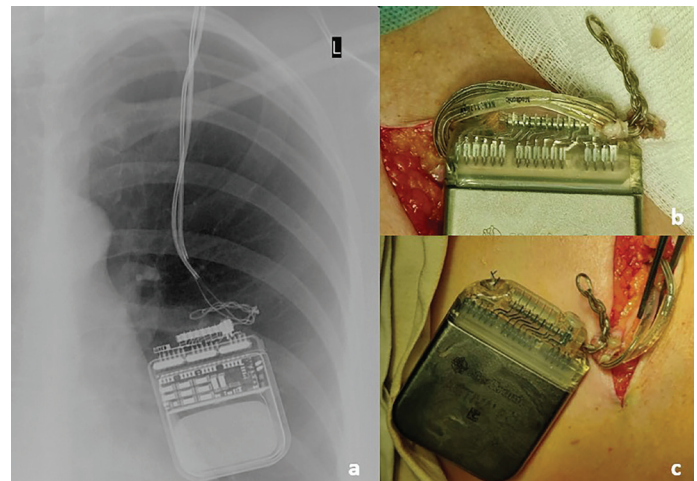


Figure 2 – Chest X-ray showing IPG and coiling of extension wires (a). Intraoperative view of the IPG and coiled of extension wires (b,c).

An urgent surgical revision was required. During the procedure, it was observed that the DBS extension leads had twisted (Figure 2b, c), necessitating replacement. Additionally, the IPG was repositioned securely beneath the pectoralis fascia to reduce the risk of future displacement and coiling.

This intervention resulted in significant improvement in the patient's dystonic symptoms, with follow-up examinations showing normalization of impedance levels.

Discussion

Deep brain stimulation is a neuromodulatory intervention that applies a neurostimulator to deliver electrical impulses to specific regions of the brain. For decades, DBS has been predominantly treating for managing motor symptoms for patients with advanced Parkinson's disease and dystonia.

While DBS provides the benefit of reversibility, it also has a few limitations. These include high costs, the requirement for continuous follow-ups, periodic battery replacements, and possible device-associated complications [9]. Among them, hardware-related complications continue as a cause for serious concern [10, 11], including Twiddler's syndrome, a rare but potentially serious complication characterized by device manipulation leading to lead displacement or fracture. Although Twiddler's syndrome has been well-documented in cardiac devices, its occurrence in DBS patients has only recently gained recognition as DBS implantations have become more common [12]. Twiddler's syndrome occurs due to excessive movement of the IPG within an oversized pocket, allowing for repetitive twisting of the device until leads are tightly coiled or displaced [1].

The condition often presents with rapid symptom recurrence, loss of stimulation efficacy, and abnormal impedance readings, leading to deterioration in motor function or exacerbation of psychiatric symptoms [6]. Additionally, Twiddler's syndrome can cause localized pain along the course of the lead wires, particularly in the postauricular region [13].

Various risk factors have been associated with Twiddler's syndrome, including psychiatric conditions such as obsessive-compulsive disorder, anxiety, and paranoia, as well as physical factors like weight loss, female sex, advanced age, and excessive subcutaneous fat reduction in previously obese individuals [14–16].

Additionally, younger patients with high mobility and repetitive motor behaviors may be predisposed to Twiddler's syndrome due to inadvertent lead twisting [17]. However, our cases did not present with behavioral disorders, high BMI, or significant physical activity, suggesting alternative mechanisms for lead displacement.

Radiographic screening plays a crucial role in early detection, with studies showing that plain X-rays can reveal twisted extension wires in most cases [4]. In our cases, early X-ray imaging enabled prompt identification and surgical intervention. All patients with Twiddler's syndrome-related hardware malfunction underwent revision surgery, underscoring the importance of timely diagnosis and intervention.

Preventative measures include surgical techniques to secure the IPG, such as fixating the device within a tight-fitting subcutaneous pocket using nonabsorbable sutures or anchoring it to the pectoralis fascia [15, 16]. Limiting pocket size and ensuring adequate closure of the pseudocapsule around the IPG may further minimize the risk of rotation [18]. The TYRX™ Absorbable Antibacterial Envelope has also been proposed as a protective measure against device migration [5]. In some cases, subpectoral or subfascial placement of the IPG within polyester

pouches has been suggested, however, these methods may not entirely prevent manipulation [13].

A potential solution to eliminating these complications is the development of a wireless IPG, eliminating the risk of lead twisting. On the other hand, making IPGs that are specific to each person's anatomy might help them fit better in the subcutaneous pocket, limiting movement and the chance of displacement.

Looking ahead, targeted gene therapy may provide a device-free solution for treating movement disorders. This field is already advancing, with multiple Phase I and early Phase II clinical trials underway. These early studies have played a crucial role in shaping the next generation of gene therapy trials, which continue to evolve and hold significant promise for more effective and long-lasting treatments [19].

Currently, due to the unpredictable nature of Twiddler's syndrome, routine follow-up with screening X-rays during the initial postoperative period is recommended to enable early detection before complications occur. Additionally, patients should be advised on activity restrictions during this period to minimize mechanical stress on the system and support stable encapsulation of the IPG [13].

Emerging evidence suggests that genetic and neurobiological factors may influence susceptibility to hardware-related complications in DBS. Genetic predispositions related to foreign body response, wound healing, and compulsive behaviors associated with neurotransmitter signaling may contribute to Twiddler's syndrome risk [8, 17]. Understanding these factors may allow for personalized DBS strategies, including patient-specific surgical techniques and postoperative monitoring plans.

Conclusion

Twiddler's syndrome is a rare but serious complication of DBS, leading to hardware failure, symptom recurrence, and the need for surgical revision. Our study identified two cases, emphasizing the importance of early diagnosis through radiographic imaging and impedance abnormalities. Preventive strategies, such as secure fixation of the IPG and careful pocket sizing, are crucial in reducing risk.

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