

## Case Report

# Schizophrenia as a Risk Factor for Pacemaker Failure due to Twiddler's Syndrome

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## Abstract

Twiddler's syndrome is rare, and thus it is remarkable to diagnose it twice in a short time in independent patients and to notice that both patients had paranoid schizophrenia. This strongly indicates a connection between Twiddler's syndrome and schizophrenia, and it is important to be aware of Twiddler's syndrome as a cause of pacemaker-failure especially in patients with schizophrenia or other mental disorders. Furthermore, it is important to inform the patient, caregivers and psychiatrists about Twiddler's syndrome with the purpose to prevent the patient from manipulating the pacemaker.

**Keywords:** Pacemaker generator manipulation; Pacemaker failure; Twiddler's syndrome; Schizophrenia

## Introduction

Twiddler's syndrome refers to a patient manipulating the pacemaker, rotating the pulse generator thereby coiling and twisting the electrode (s) and retracting the electrode (s) from the intracardiac position. This may result in pacemaker failure and lead to potentially fatal consequences [1-9].

In the Department of Cardiology, Roskilde, Denmark, about 850 pacemakers are implanted a year. By and large, there is an average of one case of Twiddler's syndrome a year. A few months apart, two cases of Twiddler's syndrome were encountered, both involving patients with paranoid schizophrenia. This points to a significant correlation between schizophrenia and Twiddler's syndrome; also, the literature supports increased incidence for patients with psychological maladaptation and mental disabilities like obsessive-compulsive disorder, bipolar disorder, depression, dementia, and developmental delay [4]. Additional risk factors are laxity, obesity, old age, female gender, inadequate fixation of a transvenous lead at the site of introduction, and a subcutaneous pocket of a capacious size allowing the pulse generator to move, perhaps due to repeated arm movements [1,2,4-9,11,12]. The situation usually occurs within the first year of implantation [2-4,7].

## Case Presentation

### Case 1

An 81-year old female was admitted to hospital from her nursing home due to bradycardia and dyspnea on light exertion (chronotropic incompetence). No syncope. ECG showed a second-degree atrioventricular 2:1 block, frequency 40, Right Bundle Branch Block (RBBB) and Left Anterior Hemiblock (LAH). She had previously exhibited intermittent Left Bundle Branch Block (LBBB).

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Echocardiography: EF 45% to 50%. Height 163, weight 95 kg.

Prior to this, she was afflicted with substituted myxedema, treated with Eltroxin 100 µg daily, and stable mental disease, treated with tablet Duloxetine 60 mg daily, tablet Quetiapine 50 mg daily, intramuscular depot Risperdal 37.5 mg every two weeks, and tablet Lysantin 50 mg pro necessitate.

Now a DDD-pacemaker was implanted. Two screw-in electrodes were introduced via the left cephalic vein; the tips were positioned in the right atrium, in the appendage, and in the right ventricle, in the distal septum. Both electrodes were introduced in ample length (Figure 1A). They were secured with non-absorbable sutures on the lead sleeves at the site of introduction. The pulse generator was positioned in place in a subcutaneous pocket and secured with a non-absorbable suture. The incision was closed with absorbable ligatures and covered with a band aid; and with a compressive bandage for 8 hours postoperatively. The pacemaker was tested during surgery and 7 hours later with excellent values, and atrial guided 100% pacing in the ventricle throughout. There were no complications, and 10 hours after the operation she was discharged to the nursing home.

Ten days later, she went to see her psychiatrist for a routine visit and the psychiatrist concluded that she had been suffering from schizophrenia, paranoid type, for many years. She had hallucinations, paranoid delusions, and recurrent abdominal sensations, "unpleasant abdominal thoughts". Previously, she had also been diagnosed with depression and dementia. Her mental condition had been relatively stable for the past couple of years and she spent most of her time knitting.

Three months later, she returned for a scheduled test of the pacemaker. The atrial electrode was functioning as it should. The ventricular electrode could neither sense nor pace, and she had a second-degree atrioventricular block. X-ray of the thorax was characteristic of Twiddler's syndrome, the Reel syndrome variant (the electrodes were not twisted, but nicely coiled around the pulse generator) (Figures 1B and C) [7]. Echocardiography and blood tests were normal.

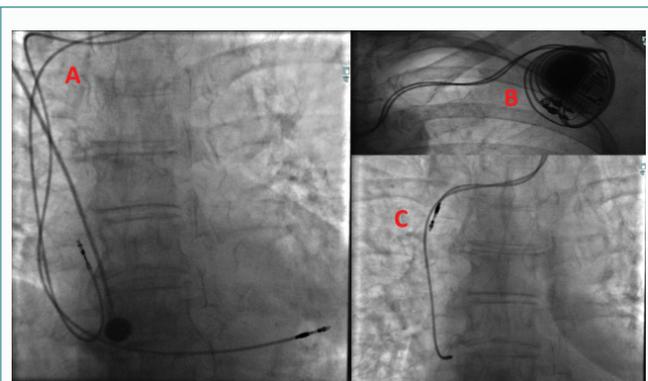
The patient had talked about nervousness and abdominal turmoil, and as a result of this, her general practitioner had administered sedatives to her. She was also given massage against back pain. Since

the pacemaker had felt inconvenient to her, she had applied Voltaren gel to the pocket area. She had not been aware of any movements of the generator. Despite the bradycardia, her general condition was good and she felt well.

At reoperation, there was no sign of electrode damage. The electrodes were replaced and safely secured, as was the pulse generator.

The patient and her family were informed about Twiddler's syndrome and advised against manipulating the pacemaker.

Since that time, there have been no problems related to the pacemaker.



**Figure 1:** (A) Screw-in electrodes of ample length were introduced at the primary pacemaker implantation procedure. (B) Three months later, the electrodes had been coiled around the pulse generator and retracted (Reel syndrome) (C) the ventricular electrode to the vena cava superior; the tip of the atrial electrode, however, remained in position in the atrium.

## Case 2

A 62-year old male was admitted from his home where he was found unconscious and with grand mal. In the emergency room he presented with a Glasgow Coma Scale of 5, and frequent and shallow respiration. ECG showed a third degree atrioventricular block and a ventricular escape rhythm with LBBB and 28 bpm. Prior to this, he had bifascicular block (RBBB and LAH). He was stabilized with Zoll pacing, atropine, and isoprenaline and transferred by ambulance to the pacemaker implanting hospital. Echocardiography was normal. He exhibited slightly elevated infectious parameters but normal temperature. Height 170 cm, weight 46 kg.

A VVI-pacemaker was now implanted in the left pectoral region. A screw-in electrode was introduced into the right ventricle via the cephalic vein and secured on the sleeve to the fascia. The pulse generator was positioned and secured in the subcutaneous pocket. The compressive bandage was removed after a few hours. The entire procedure was without complications and the test values remained stable.

Initially, motoric unrest and abrupt movements called for sedative treatment and constant observation. His condition improved slowly, and after a few days, he had regained his usual kind although conspicuous behavior.

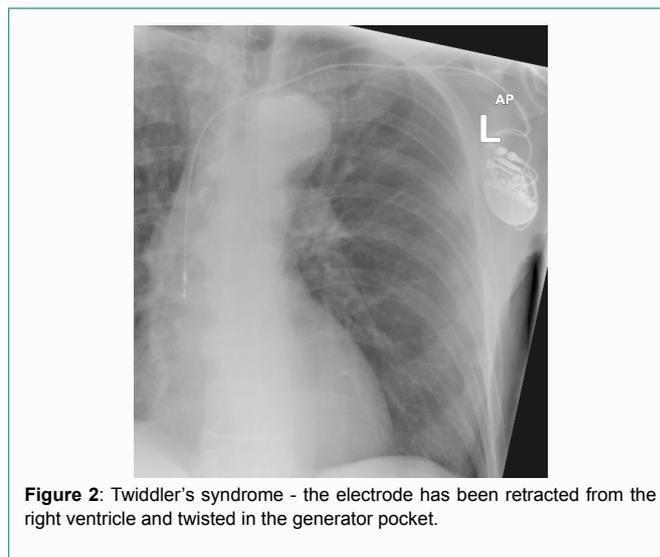
This patient had been suffering from schizophrenia, paranoid type, since his youth and had been on disability pension since the age of 30. In addition to this, he had been having a drinking problem for many years further complicated by neuropathy, Charcot joints, wounds, and osteitis. During his hospital stay, it was evident that he had difficulties

with his balance and also had cognitive deficits and was in massive need of help. He was treated with Truxal 15 mg  $\times$  4, Anafranil R 75 mg  $\times$  2, Trilafon decanoate 160 mg intramuscular injections every other week, Campral 333 mg  $\times$  3, and Antabuse 800 mg  $\times$  2/week.

He looked miserable and had infected wounds, especially on his toes and feet. *Streptococcus haemolyticus* was found in his blood and in his wounds. He was administered antibiotics, his wounds were cleaned and he was discharged 10 days after the pacemaker implantation to a nursing home.

One week later, he was readmitted due to grand mal, unconsciousness and a high fever. He had infected wounds again and a septic knee arthritis. X-ray of the thorax showed a retracted and twisted pacing electrode (Figure 2), characteristic of Twiddler's syndrome. ECG showed sinus rhythm, normal PQ, frequency 100, RBBB and LAH, and pace spikes revealing lack of sense and lack of capture.

The existing pacing system was removed; no signs of local infection were observed. Because of the widespread and serious infections and neurologic reactions, he had a temporary pacemaker implanted. More than two months went by with complicated treatments and slow improvement in the patient's condition before it was safe to implant a permanent pacemaker. This procedure did not present any complications, and no problems with the pacemaker have been observed since then.



**Figure 2:** Twiddler's syndrome - the electrode has been retracted from the right ventricle and twisted in the generator pocket.

## Discussion

The typical patient with Twiddler's syndrome is an old woman, obese, with tissue laxity, and a mental disturbance [4], as in Case 1 presented above. Generally, this is a rare complication. However, the condition is observed more frequently in patients with mental disorders, where it has to be taken into account as a cause of electrode displacement, pacemaker malfunction or pacemaker failure. The present case stories indicate that schizophrenia pose as a strong risk factor (Cases 1 and 2).

Twiddler's syndrome results when a patient - intentionally or unintentionally - manipulates (rotates) a device, most commonly seen with implanted cardiac pacemakers of all types, but it can also occur with chest ports, deep brain stimulators, and other devices

[3,4,8,9,12]. Localized discomfort may predispose the patient to manipulate the device [11].

Pacing follows the retracted lead. Retraction may result in loss of sensing and loss of cardiac pacing. In a pacing dependent patient, bradycardia may cause dizziness and syncope and asystole may be lethal [2,3,7]. In a patient with an ICD, there is a risk of inappropriate shocks [2,3,12]. Instead, pacing stimulates nearby structures, such as the phrenic nerve, causing contraction of the right hemidiaphragm, sensation of abdominal pulsations, hiccups, involuntary inspirations, and dysphonia; and the brachial plexus causing rhythmic arm twitching [1,2,4-6,8,9].

Chest X-ray is diagnostic. It shows twisting or coiling and displacement of the electrode(s) (Figures 1 and 2) [1,2,6-9], sometimes insulation leakage or lead fracture, and a change in position of the generator [4,11]. ECG and pacemaker interrogation may show loss of sense and loss of capture [3,4,7-9].

The treatment involves revision, re-positioning of the electrode (s), if damaged (typically when twisted) a new electrode, suture fixation of the lead sleeve, minimizing the pocket size, securing the device to the underlying fascia with a non-absorbable ligature [4,8], or securing it in a tight, sub muscular pocket [1,3,7]. Postoperatively firm compression is advised to prevent fluid accumulation and thereby enlargement of the pocket [1,7].

Before performing a pacemaker implantation, the cardiologist should be aware of the patient's risk factors and consider technical solutions, including sub muscular implantation or a leadless pacemaker [1-4,6,7,9,12].

It is surprising, that Twiddler's syndrome developed in these two cases of correctly performed primary implantations, with fixation of electrodes and pulse generators. It seems as if patients with paranoid schizophrenia are able to resist the pain caused by manipulation of the pace electrodes and pulse generators [10]. This emphasizes the importance of advising against twiddling the generator [2,4], and carefully informing the patient and caregivers about the nature of the action of the pacemaker and what to do in the event of suspected pacemaker failure [3,10].

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