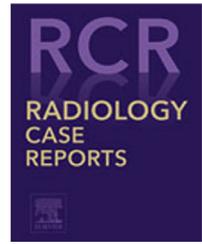


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## Case Report

# Twiddler's syndrome: Between mechanical recoil and behavioral troubles ☆,☆☆

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### ABSTRACT

A 72-year-old male with severe dilated cardiomyopathy, benefited of implantable cardiac defibrillator implementation. Device control shows high impedance. On X-ray, electrodes were completely twisted in the generator pocket, they were replaced and the generator was fixed to pectoralis-major fascia. Nurses report patient abnormal movements, scratching implantation area. This was identified as probably the trigger of the complication.

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## Introduction

Twiddler's syndrome (TS) was first described in 1968 by Bayliss et al. [1] in patients with pacemakers. It consists of the rotation of the generator by its axis, which causes the torsion electrodes. This torque may lead to fracture or displacement of electrodes, causing dysfunction device. It has also been described in patients with implantable defibrillator [2]. We present a case of a patient with this complication in which a precipitating factor could be identified in an old man with behavior troubles.

## Case report

This is a 72-year-old man with 6 years history of worsening exercise intolerance compatible with heart failure in relation to a severe dilated cardiomyopathy, who was initially admitted for cardiac decompensation. In the subsequent study, coronary angiography found nonsignificant irregularities in the coronary arteries and left ventricular ejection fraction was at 28%. We decide to set up a double chamber defibrillator for primary prevention of sudden cardiac death (SCD) which was performed without complications. At follow-up, the patient

Abbreviations: TS, Twiddler's syndrome; DCM, dilated cardiomyopathy; SCD, sudden cardiac death; ICD, implantable cardiac defibrillators; IPG, internal pulse generator.

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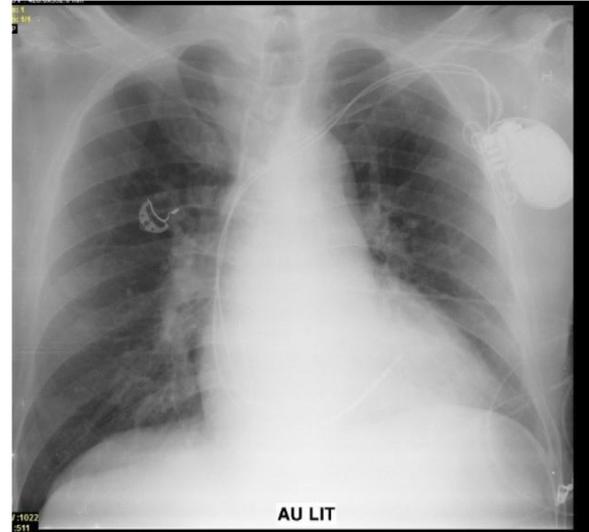


**Fig. 1 – Chest X-ray showing the coiling of the device leads around the device in a 72-year-old man.**

remained stable without ventricular arrhythmias or inappropriate discharges of the defibrillator. Periodic controls parameters were always correct.

Two months later, he was admitted to emergency department for lipothymia. Also, family reports onset of behavioral disorders. Interrogation of implanted device showed normal functions without recorded tachycardia episodes, however.

Investigations found a recent frontal ischemic stroke. Patient was discharged with a neuropsychiatric follow-up. One month later, the control of the device shows a high impedance of electrode stimulation above 2000  $\Omega$  (ohm), suggesting a fracture thereof.



**Fig. 3 – Chest X-ray control after replacement of new leads.**

In the chest X-ray, electrodes were completely twisted in the generator bag, with the traction of the intracavitary electrode (Fig. 1).

Electrode extraction was carried out by continuous traction (Fig. 2), and others were replaced, chest X-ray control was done (Fig. 3). Also, the generator was fixed to the fascia of the pectoralis major.

In the days after the intervention, the nurses report repetitive abnormal movements of patient touching and scratching the device implantation area, he unconsciously repeats these stereotyped movements. This movement was identified as probably trigger of the complication, so the patient was advised to avoid this maneuver in the future.



**Fig. 2 – Operative view of the extraction of the twisted leads of the ICD.**

The patient was integrated to a cardiac rehabilitation program with neuropsychiatric and cardiological follow-up.

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## Discussion

TS is a rare mechanical cause of permanent pacemaker's dysfunction. The frequency would vary between 0.07% and 7%.

Firstly described by Bayliss in 1968, it was defined as manipulation of the cardiac implantable device around its central axis within skin pocket which causes the coiling and dislodgment of electrodes [1,3]. This phenomenon usually occurs in the first year following device's implantation; the earliest case described on the pacemaker had a delay of 17 hours after implantation [4]. More recently, a late Twiddler syndrome has also been described [5].

Furthermore, cases of similar syndrome have been reported with implantable cardioverter-defibrillators and cardiac resynchronization therapy [6,7]. In fact, the first case of Twiddler's syndrome associated with automatic implantable defibrillators (ICD) was described by Veltri et al. in 1984 [2].

The main risk factors predisposing to the development of TS are: advanced age, female sex, obesity, cognitive, and behavioral disorders. In the elderly, the increased laxity of the subcutaneous tissues favors the displacement of the material, also the creation of too large pockets for internal pulse generator (IPG) facilitates their rotation into the skin pocket most often during physical activities [4,8,9]. In some isolated cases, it has been possible to demonstrate active manipulation of the generator by the patient [10].

Otherwise, a recently published series of cases have suggested that the construction of an implantable pulse generator (IPG) may itself be a predisposing factor: the presence of a single anchoring hole intended for attachment exposes more to displacement of the IPG [11].

Clinically, the displacement of the device is itself painless, the voluntary or unintentional manipulation is often denied by the majority of patient. TS is characterized by the reappearance of neurological or cardiac disorders attributed to the pacemaker dysfunction in dependent patients, which can have dangerous consequences. In our case, the dysfunction is totally asymptomatic but the consequences could be fatal for the patient.

The other clinical manifestations, displacement or fracture of the electrodes can cause minor complications, such as increases in impedance or stimulation threshold [12], or stimulation of the skeletal muscles, but there may also be major complications, such as failures in under or overdrive, as well as increases in the defibrillation threshold [13].

Positive diagnosis of this syndrome is very simple; it's based on the chest X-ray which shows the displacement of the device with rotation of the conductive wires [7].

Therapeutic procedure is based on the uncoiling of the electrodes, the implantation of new leads with repositioning and fixation of the device [7,9,14]. Some authors encourage the reutilization of the same electrodes.

Active fixations of leads and suturing of the device to the fascia can also help prevent displacement [1,15]; some teams

reserve this option for patients with mental disorders and elderly ones with loose subcutaneous tissues [4]. We can also implant it below the pectoral muscle [16], that's what we did in our patient by the use of a Dacron patch, which promotes tissue growth around the device and ensures better fixation. Also, minimizing the pocket size without redundant space around the generator is essential to prevent the development of this syndrome [4,5].

In addition to optimizing fixation procedures, the education of patients, especially the elderly, is one of the most important means of avoiding manipulation of the device [7].

Radiological follow-up of predisposed patients may also help to make an early diagnosis and thus avoid major complications.

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## Conclusion

Twiddler's syndrome is currently recognized as an uncommon mechanical cause of intracardiac device malfunction, especially in the elderly. Its diagnosis is very easy based on chest X-ray. Optimization of the device fixing measures and educating patients to avoid any manipulation of the pulse generator are the most important methods of prevention.

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## Patient consent

Patient consent was obtained from the patient.

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