

TWIDDLER SYNDROME AND ITS VARIANTS (REEL SD AND RATCHET SD)

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SUMMARY

Twiddler syndrome is secondary to deliberate or involuntary manipulation of the box by the patient. The axis of rotation is sagittal, causing winding, damage and displacement of the leads. This is a descriptive retrospective study of a series of 3 cases of Twiddler syndrome and its variants. Carried out over a period of 2 years and 5 months, within the cardiology department B, in order to specify the predictive factors, the time to onset, the means of diagnosis as well as the therapeutic modalities of these syndromes. The treatment includes fixation of the box with replacement of the leads in Twiddler syndrome and repositioning in Reel syndrome. Twiddler syndrome and its variants are rare but potentially lethal mechanical complications of implanted cardiac pacemakers and defibrillators. Educating the patient on the importance of never handling the box remains the best method of prevention.

KEYWORDS: Pacemaker, Box, manipulation, Twiddler syndrome, Reel, Ratchet.

INTRODUCTION

Since the 1950s, cardiac stimulation has evolved, both in its indications and on a technical level. Despite biotechnological advances, complications related to cardiac pacemakers remain constant.

These complications can occur

- **Perioperatively:** immediate vascular or cardiac problems.
- **Early postoperative:** local or hardware-related complications.
- **Late:** endocarditis, probe fractures, late displacements, chronic venous occlusions.

Lead displacement is common and can occur early or late, leading to ECG disturbances and symptoms in pacemaker-dependent patients. Diagnosis is based on ECG, chest X-ray, and device interrogation, sometimes requiring reintervention to reposition the lead.^[1,2,3]

Twiddler syndrome and its variants, although rare, are serious, with leads that may be completely withdrawn or retracted into the casing. These cases require specific management to prevent potentially life-threatening complications.^[1,2]

MATERIALS AND METHODS

The objective of our study is to specify the predictive factors, the time to onset, the means of diagnosis as well as the therapeutic modalities of Twiddler syndrome and its variants. This study is presented in the form of a

descriptive retrospective study on a series of 3 cases of Twiddler syndrome and its variants. The data were collected at the Cardiology Department B of the Souissi Maternity Hospital in Rabat over a period of 2 years and 5 months, from January 1, 2021 to May 30, 2023.

The study population included all patients hospitalized for pacemaker lead displacement during the study period. Inclusion criteria included patients hospitalized for management of Twiddler syndrome and its variants, namely Ratchet syndrome and Reel syndrome. Exclusion criteria excluded patients admitted to the department for a complication other than lead displacement, as well as those admitted for lead displacement other than Twiddler syndrome and its variants.

RESULTS

	Observation 1	Observation 2	Observation 3
Sexe / Age	Femme / 67 ans	Femme / 80 ans	Femme / 75 ans
FRCVX modifiables	HTA/ Obésité	DT2 / Obésité	HTA
ATCD	PM double chambre	PM mono chambre	<ul style="list-style-type: none"> PM double chambre Démence
Délais avant le Dc	1 mois	1 année	3 mois
Motif	<ul style="list-style-type: none"> Contactions rythmique bu bras droit Malaise lipothymique 	<ul style="list-style-type: none"> Syncope 	<ul style="list-style-type: none"> Découverte fortuite lors de la consultation de contrôle (interrogation du PM et ECG)
Examen clinique	<ul style="list-style-type: none"> Lésions de grattage en regard de la loge du boitier Bradycardie 	<ul style="list-style-type: none"> Bardycardie 	<ul style="list-style-type: none"> Bardycardie
ECG	BAV 2/1 à QRS fins et CVM à 36bpm	un BAV complet à ORS larges et CVM à 40bpm avec des spikes ventriculaires révélant à la fois un défaut de capture et de détection inappropriés	BAV complet à QRS fins et CVM à 38bpm
Interrogation du Pacemaker	Augmentation brusque de l'impédance de stimulation des deux sondes	—	Augmentation de l'impédance de stimulation des deux sondes
Rx thorax	Sondes déplacées, tordues et complètement enroulées autour du boitier et entre elles	Sonde déplacée et complètement enroulée autour et derrière le boitier	Sondes déplacées, tordues et enroulées l'une contre l'autre
Diagnostic	Association Sd de Twiddler et Sd de Reel	Sd de Reel	Sd de Twiddler
Traitement	<ul style="list-style-type: none"> Repositionnement et fixation des sondes Fixation du boitier 	<ul style="list-style-type: none"> Remplacement de la sonde Fixation du boitier Réduction de la loge 	<ul style="list-style-type: none"> Repositionnement et fixation des sondes

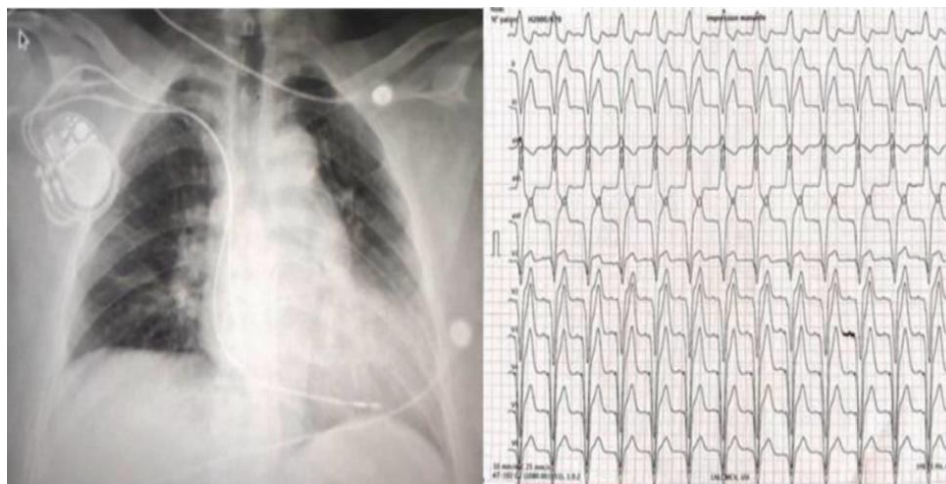


Figure 1: X-ray (probes in place) and post-operative control ECG (electro-driven rhythm)

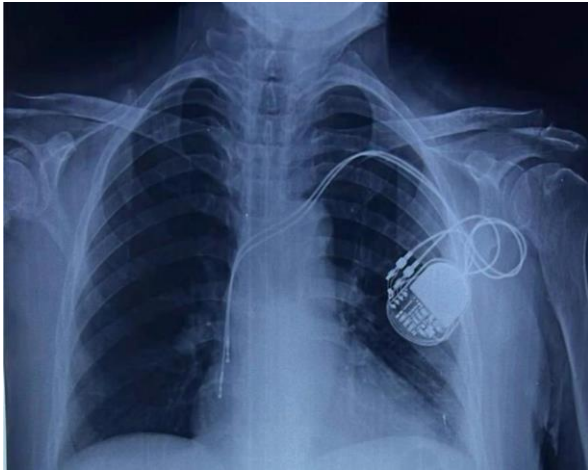


Figure 2: Chest X-ray: Probes displaced, twisted and rolled up near the housing.

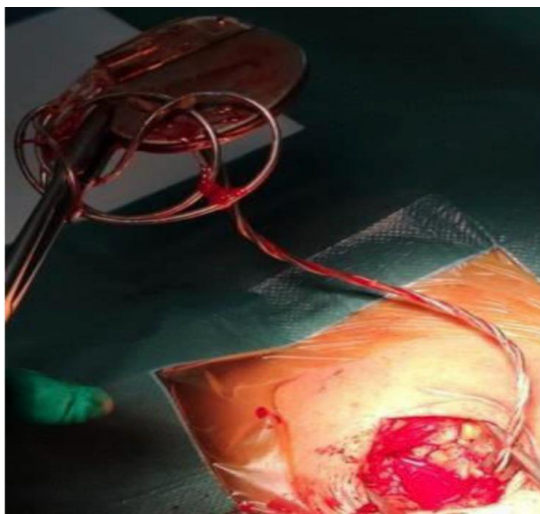
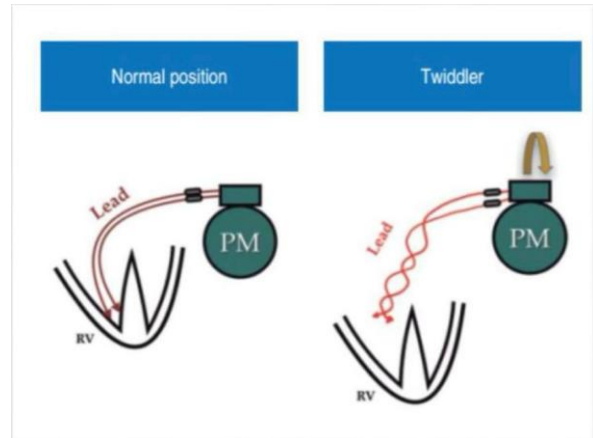


Figure 3: Intraoperative image: probes twisted and wrapped around the box.

DISCUSSION

1- DEFINITION OF TWIDDLER SYNDROME

The term "twiddler syndrome" or Twist syndrome was first described by CE Bayliss in 1968 in a patient with a pacemaker. In 1984, Veltri reported the first case in a patient with an implantable cardioverter defibrillator (ICD). Twiddler syndrome is characterized by the displacement of one or more leads due to deliberate or unconscious manipulation of the housing by the patient. This manipulation causes rotation of the housing, leading to retraction and coiling of the leads, which can lead to fracture. This syndrome affects pacemaker, implantable cardioverter defibrillator and cardiac resynchronization therapy housings.^[8,9,10,11]



Schematic diagram of the mechanism of Twiddler Syndrome.

2- The incidence

Twiddler syndrome is a rare complication of cardiac implantable electronic devices (CIEDs), with a reported incidence ranging from 0.07% to 7% across studies. Significant multicenter studies have reported incidences of 3.3% (FOLLOWPACE), 1.4% (Ghani *et al.*, 2014), and between 1% and 2.69% (Wang *et al.*, 2018). More recently, in 2022, O. Gomez *et al.* reported an incidence of 1.2%. These variations highlight the need for close patient monitoring to detect this potentially serious complication.^[1,2]

3- TIME OF APPEARANCE

The majority of cases of twiddler syndrome are diagnosed within the first year after implantation, usually over a period of several months to a year^[15], but it can occur at any time after device implantation.^[16]

Concerning our patients: in the 2 patients who were diagnosed with SD Twiddler syndrome (observations 1 and 3) the delay before DC was between 1 month and 3 months.^[11,22]

4- PHYSIOPATHOLOGY

1. Twiddler syndrome secondary to handling of the box

Twiddler syndrome typically occurs after manipulation of the implantable cardiac device casing. Patients with cognitive or psychiatric disorders, such as dementia or obsessive-compulsive disorder, are at particular risk. Even without a medical history, some patients manipulate their device in response to the presence of a foreign body, although most deny any manipulation.^[1,11,27]

2. Twiddler syndrome outside of any manipulation of the box

Some cases of Twiddler syndrome occur spontaneously, without deliberate manipulation. Several factors increase the risk of this syndrome:

- Advanced age: hyperlaxity of subcutaneous tissues.
- Female sex: weighting effect of the breasts.
- Obesity: accumulation of subcutaneous fatty tissue.

- Excessive arm movements on the housing side.
 - Sports involving repetitive arm movements (golf, swimming).
 - Reabsorbed lodge hematoma.^[16]
 - Small size of the device compared to its housing.
- In the 3 reported cases, the patients were elderly women (mean 74 years), two were obese and one had dementia.

6- Modes of revelation

Twiddler syndrome presents with a variability of symptoms, ranging from asymptomatic to severe cases that can lead to sudden death. Device malfunctions can cause varied clinical manifestations depending on the site of the displaced lead: arm twitching, vagal symptoms, or diaphragmatic contractions. These neuromuscular manifestations make diagnosis difficult, but Twiddler syndrome should be considered in any patient with a cardiac device presenting with such symptoms. In patients with implantable cardioverter-defibrillators (ICDs), the syndrome can cause inappropriate shocks or no shocks in the presence of ventricular arrhythmias.^[11,21]

Concerning our patients, two were symptomatic and in the third patient the discovery of this syndrome was fortuitous.

7- Positive diagnosis

The electrocardiogram and chest X-ray are the first-line tests for diagnosing Twiddler Syndrome. The ECG can detect sensing defects or loss of capture, while the chest X-ray reveals displacement and coiling of the leads as well as rotation of the housing. Telemetric monitoring, by interrogating the device, confirms the diagnosis by showing an increase in the stimulation impedance, a sensing defect, an increase in the stimulation threshold, or even a total loss of capture.

In our patients, ECG and chest X-ray were performed in all patients, and pacemaker interrogation was performed in only two patients.^[11,22,25]

8- Treatment

If the device malfunctions, surgical intervention is necessary. Diagnosis is confirmed if the leads are coiled. Treatment usually consists of replacing damaged leads and securing them with non-absorbable suture. Sometimes leads can be untwisted, repositioned, and secured if they are not fractured. The housing pocket is reduced for optimal fit, and fixation of the housing to the muscle fascia may be considered, especially in patients with mental disorders. The use of a Dacron pouch may help by promoting fibrous tissue growth around the housing, reducing the risk of movement, and is recommended for patients at high risk of recurrence.^[13,16,26,27]

Regarding our patients, one patient benefited from replacement of the probes with active fixation of the

probes and the box and the other two benefited from repositioning only.

9- REPEATED

Prerogative of patients with proven psychiatric illness, poorly or not treated. Who continue to manipulate their boxes.^[28]

10- Prevention

Prevention of Twiddler syndrome is based on two main aspects

1. Patient and family education: Inform about the consequences of handling the box and the importance of never touching it.

2. Identification of patients at risk: Implement preventive measures such as securing the device and leads with non-absorbable sutures during surgery, and creating a small or retropectoral pocket. Postoperatively, apply firm compression around the shoulder and chest to prevent fluid accumulation and enlargement of the pocket. Regular monitoring of device function and lead placement, by telemetry and chest X-ray, is essential for early diagnosis of the syndrome.^[11,16,29]

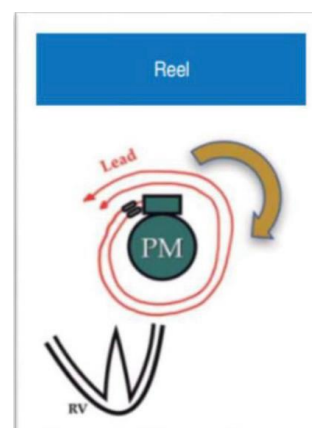
11- OTHER VARIANTS OF TWIDDLER SYNDROME

1. Revers twiddler's syndrome

Reverse Twiddler syndrome occurs when spontaneous manipulation or rotation of the can is in the opposite direction to that of classic Twiddler syndrome. This also results in coiling of the leads, but unlike Twiddler's syndrome, it causes the leads to advance toward the heart rather than retract.^[17,27]

2. Reel Syndrome

Reel syndrome is characterized by rotation of the implantable cardiac device housing along its transverse axis, in a clockwise direction. This movement causes the leads to move and wind around the housing, similar to a fishing reel, spool, or yo-yo. This syndrome was first described by Carnero-Varo in 1999 in a 70-year-old patient, one month after implantation of a single-chamber pacemaker.^[17,28]

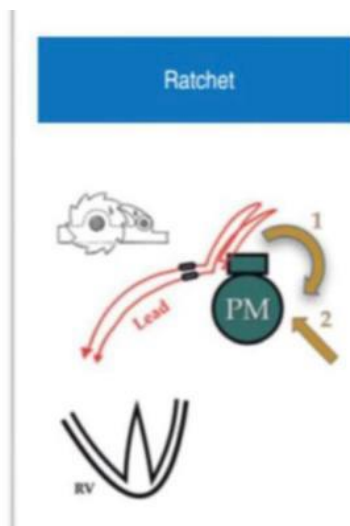


Reel Sd Schematic

3. Ratchet Syndrome

Ratchet syndrome, identified by Von Bergen in 2007, is a variant of Twiddler syndrome in which the cardiac device housing is manipulated laterally along the coronal plane, in a clockwise direction, without rotation on its axes. This action results in the detachment of the lead, which is slowly advanced through the suture sleeve like a one-way brake until it reaches the housing. This one-way withdrawal can also occur spontaneously with arm movements corresponding to the lead and insufficient lead fixation. Predisposing factors include a lateral incision, proximity to the shoulder joint, loose sutures, passively fixed leads, device/housing mismatch, and incomplete housing fixation. Increased laxity and thickness of the subcutaneous tissue in obese patients are also risk factors. In most cases, the leads are displaced but not damaged, requiring only repositioning without replacement.^[17,27,28]

In our second patient who was diagnosed with Reel Syndrome, we replaced the probe instead of repositioning it due to the fracture of the distal end of the probe observed intraoperatively.



Ratchet SD Schematic

CONCLUSION

Twiddler syndrome is a rare and potentially fatal complication of implanted cardiac pacemakers and defibrillators, often caused by manipulation of the case resulting in coiling and dislodgement of the leads, which can lead to ventricular pacing failure. It can also occur spontaneously in patients with risk factors such as obesity, older age, female gender, excessive ipsilateral arm movements, and history of compartment hematoma. Twiddler syndrome typically occurs within the first year after pacemaker implantation and has a range of clinical manifestations, ranging from no symptoms to severe complications.^[1,12,29]

Reel and Ratchet syndromes share the same predisposing factors as Twiddler syndrome, but differ in the axis of rotation of the can and the nature of the manipulation.

Chest radiography is essential for rapid diagnosis. Treatment of Twiddler syndrome involves replacement and fixation of damaged leads, whereas in Reel and Ratchet syndromes, leads are often repositioned without requiring complete replacement. Prevention relies primarily on educating patients and their caregivers about the importance of never manipulating the can to avoid these potentially serious complications.^[17,26,28]

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