

## Serial Case of Twiddler Syndrome

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**Background:** Twiddler syndrome is an infrequent but potentially dangerous complication of device therapy for dysrhythmias. This syndrome results from manipulation of implanted pulse generator by the patient, leading to traction and subsequent lead dislodgement. It can also occur spontaneously. It has been increasingly reported with pacemaker or implantable cardioverter-defibrillators (ICDs). In this reports, we describe two patients with Twiddler syndrome with substantial retraction of their lead who denied any manipulation of their device.

**Case Illustration:** The first patient was a 56 year-old man with single-chamber ICD due to dilated cardiomyopathy (DCM) with congestive heart failure and severe systolic left ventricular dysfunction (ejection fraction 18%). The dislodged lead causing rhythmical twitching of left pectoral muscles and abdominal pulsations. The second patient was a 69 year-old man with dual-chamber pacemaker due to total atrioventricular block with normal systolic left ventricular function (ejection fraction 70%). It manifested as dyspnea on effort, and he also underwent pacemaker implantation. They underwent primary devices implantation at April 2016 and reposition of generators and its leads in December 2016. The first and second patients denied of manipulating the generator of ICD or pacemaker and rotated their left arm and right arm, respectively, after implantation.

**Summary:** Other unconscious arm abduction during sleep or increased muscular activity of the shoulder and arm might have led to repetitive motions within the pocket and dislodge the device. Adequate individualized patient and family education and regular evaluation every 6 month of the leads position with fluoroscopy or chest X-ray is advisable.

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**Keywords:** Twiddler syndrome, implanted cardioverter-defibrillator (ICD), dual chamber pacemaker

## Laporan Dua Kasus Sindrom Twiddler pada Pasien dengan Pemasangan *Implanted Cardioverter-Defibrillator* dan *Permanent Pacemaker*

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**Latar belakang:** Sindrom Twiddler merupakan komplikasi yang jarang terjadi namun berpotensi bahaya pada tatalaksana disritmia. Sindrom ini terjadi akibat manipulasi pada generator yang menyebabkan tarikan sehingga *lead* terlepas atau terjadi spontan. Komplikasi ini semakin banyak dilaporkan pada pasien dengan *permanent pacemaker* (PPM) atau *implanted cardioverter-defibrillator* (ICD). Laporan kasus ini menunjukkan dua pasien dengan ICD dan PPM yang mengalami sindrom Twiddler walaupun kedua pasien tersebut menyangkal melakukan manipulasi pada alat.

**Ilustrasi kasus:** Pasien pertama, seorang laki-laki berusia 56 tahun dengan *single chamber* ICD karena kardiomiopati dilatasi (DCM) dengan fraksi ejeksi 18%. Keluhan sindrom Twiddler dirasakan sebagai kejutan listrik yang teratur di dinding dada dan pulsasi di abdomen. Pasien kedua seorang laki-laki 69 tahun dengan *dual chamber pacemaker* karena penyumbatan atrio-ventrikular total (TAVB) dengan fungsi ventrikel kiri normal. Manifestasi sindrom ini adalah pasien merasakan keluhan yang sama seperti sebelum pemasangan *pacemaker* yaitu sesak saat beraktivitas. Kedua pasien menjalani implantasi alat pada April 2016 dan reposisi dilakukan pada Desember 2016. Keduanya sama-sama menyangkal melakukan manipulasi pada alat.

**Kesimpulan:** Gerakan yang tidak disadari saat tidur dan peningkatan aktivitas otot bahu dan lengan menimbulkan gerakan berulang yang memicu tarikan pada *lead* sehingga terjadilah sindrom Twiddler. Edukasi pada pasien dan keluarga serta evaluasi teratur dengan fluoroskopi atau rontgen thorax secara berkala setiap enam bulan sangat diperlukan.

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**Kata kunci:** sindrom Twiddler, *implanted cardioverter-defibrillator* (ICD), *permanent pacemaker* (PPM)

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### Introduction

Twiddler syndrome is an uncommon complication of device implantation with a frequency of 0.07–7%.<sup>1,4</sup> It is an infrequent but potentially dangerous complication of device therapy for dysrhythmias. This syndrome results from the manipulation of the implanted pulse generator by the patient, leading to traction and subsequent lead dislodgement. Originally described by Bayliss as a complication of pacemaker implantation, it has been

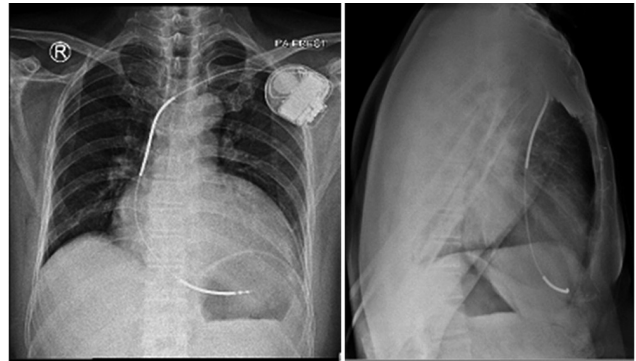
increasingly reported with implantable cardioverter-defibrillators (ICDs).<sup>1,4</sup> In this report, we describe two patients with Twiddler syndrome in an ICD and dual chamber pacemaker with substantial retraction of leads who denied any manipulation of their devices. They underwent reposition of generators and their leads in December 2016.

### First Case

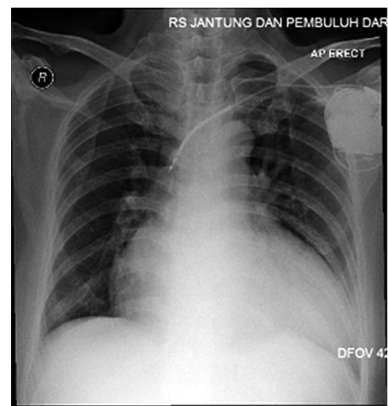
A 56-year-old man (height 172 cm, weight 76 kg) with a history of congestive heart failure due to dilated cardiomyopathy, a left ventricular ejection fraction of 18%, underwent an ICD implantation. The ICD was Medtronic D264VRM SN, PVZ6084125 (Medtronic Inc., Minneapolis, Minnesota) and the lead was Medtronic MR conditional 6947m-62cm SN TDK179870V (Medtronic Inc.). Appropriate sensing, pacing, and defibrillation thresholds were obtained at implantation. A chest X-ray obtained immediately after implantation demonstrated ideal lead position (Figure 1). Patient had no current or history of psychiatric disease.

Eight-months after ICD implantation, patient's wife saw repetitive twitching of the left major pectoral muscle and abdominal pulsations. Patient denied to have manipulated the device. He often rode a bicycle as a physical activity, but his wife said that he sometimes lifted it. He was just retired as a journalist for a year and physically active. He drove motorcycle for his daily activities. A chest X-ray revealed marked retraction of the lead, being withdrawn from the subclavian vein with its tip now positioned over the major pectoral

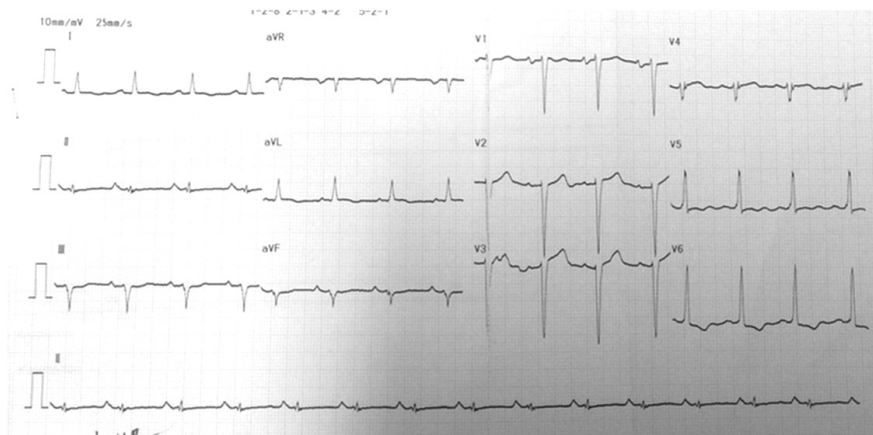
muscle (Figure 2). The electrocardiography showed sinus rhythm with 90 beat per minute (bpm) heart rate (Figure 3).



**Figure 1.** Posteroanterior and lateral chest X-Ray demonstrating appropriate lead placement in the right ventricular apex.

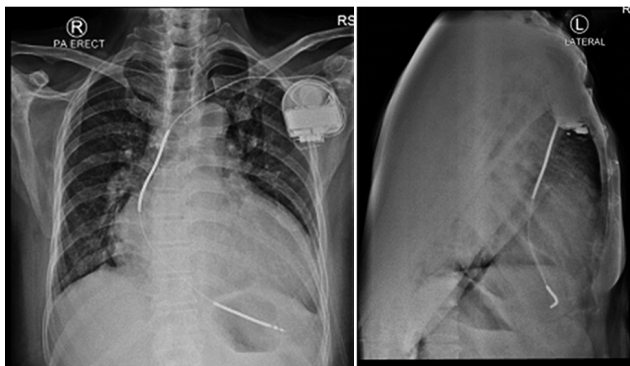


**Figure 2.** Posteroanterior chest X-Ray shows substantial retraction of the lead.



**Figure 3.** ECG shows sinus rhythm (before reposition of ICD).

The patient subsequently underwent reposition of the ICD in left deltopectoral junction area and its lead into right ventricle (RV) apex. The lead was fixated to the surrounding muscle and tissue. An appropriate size of generator pocket was tailored, the generator then implanted to the pocket. At twenty-four hours after reposition ICD and the lead, there has been no further “twiddling” (**Figure 4**) and the electrocardiography showed sinus rhythm with 70 bpm heart rate (**Figure 5**).

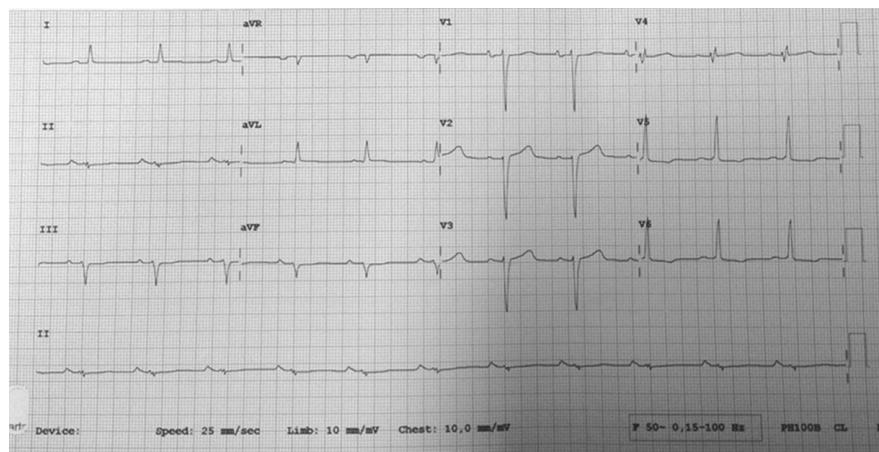


**Figure 4.** Posteroanterior and lateral chest X-Ray (post reposition of lead and ICD).

left ventricular ejection fraction of 70% and wall motion of global normokinetic. He presented in a haemodynamically stable condition without a visible pulsating abdominal mass or gastrointestinal complaint. The peripheral pulsations were symmetrically intact. His electrocardiography (ECG) showed both atrial and ventricular spikes uncaptured and his own rhythm back to marked total atrioventricular block with junctional rhythm (**Figure 6**). A chest X-ray revealed that both leads had been dislodged. Atrial and ventricular lead was coiling several times around the generator (**Figure 7**) and the diagnosis of Twiddler syndrome was made.

The patient denied in manipulating the pacemaker and rotating his right arm after implanted pacemaker. Although retired, he was still actively assisting his wife to go to the traditional market. He drove the motorcycle almost every day, a month after pacemaker implantation. Almost all of those daily activities had to be managed by his right arm. The patient had neither current nor history of psychiatric disease and can follow direction, but his wife sometimes saw unconscious arm abduction when he was asleep.

We performed to reposition of leads and generator

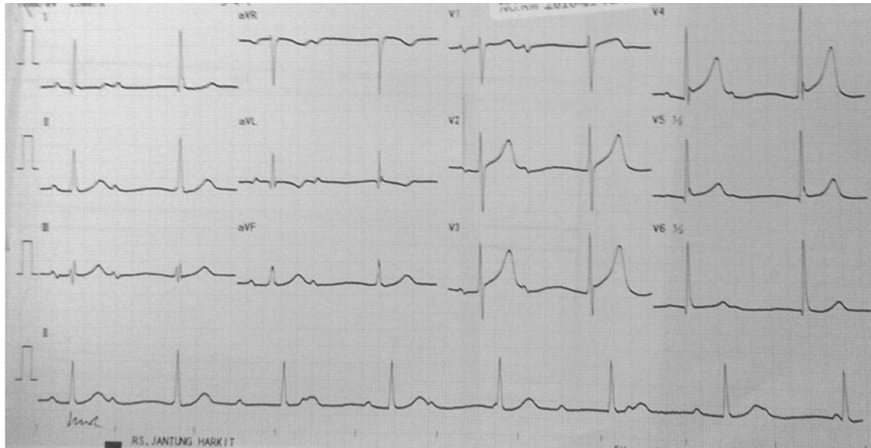


**Figure 5.** ECG shows sinus rhythm (post reposition of lead and ICD).

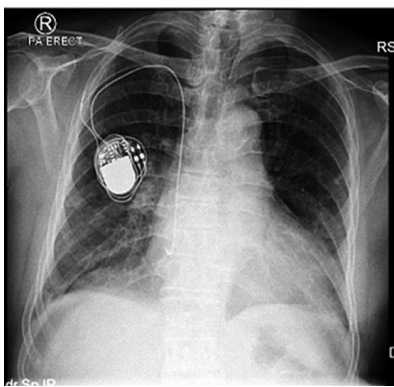
## Second Case

A 69 year-old man (height 160 cm, weight 70 kg) was referred to our hospital from Zainul Abidin Hospital Aceh because of total atrioventricular block (TAVB) on pacemaker. His medical history reported a DDD pacemaker (Medtronic Sensia 5076) implantation eight months ago because of TAVB with

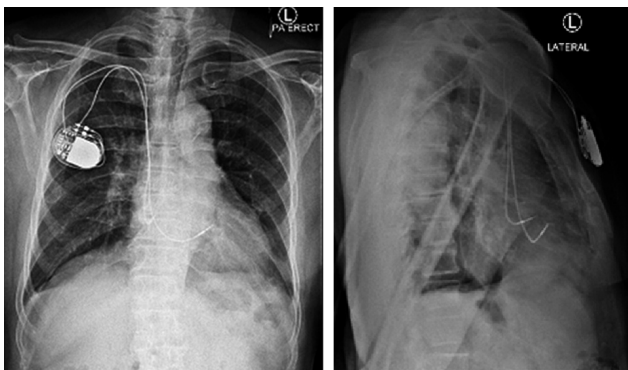
pacemaker. The atrial lead was found in right subclavian vein and ventricular lead was found in right atrium. The right atrial lead was repositioned and placed at the right atrial appendage and the right ventricular lead at the right ventricular outflow tract (RVOT) (**Figure 8**). Both leads were fixated with non-absorbable suture on its sleeve with surrounding fascia and muscles. The pocket was made within



**Figure 6.** The electrocardiography showed total atrioventricular block with junctional rhythm.



**Figure 7.** Posteroanterior chest X-ray showing substantial retraction of the lead.



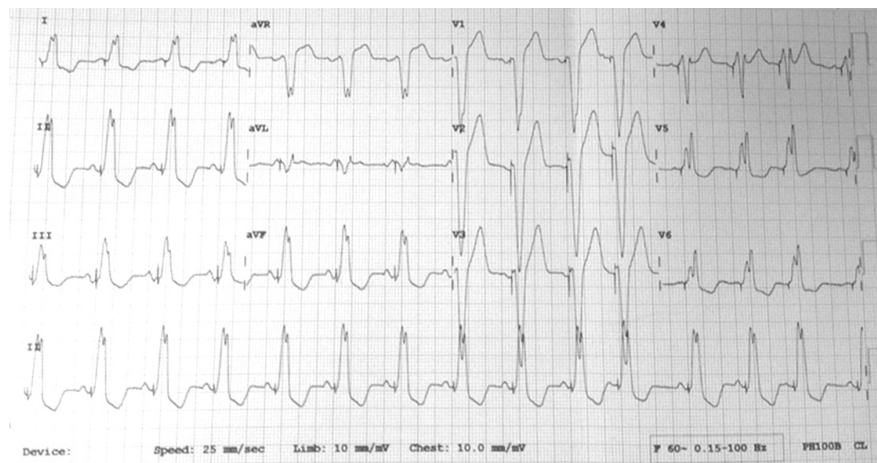
**Figure 8.** Posteroanterior and Lateral chest X-ray (post reposition of lead and generator pacemaker).

the sub-pectoral muscle for adequate fixation. After subsequent follow up, there was no complications have been noticed. The electrocardiography a day after reposition pacemaker and its leads showed atrial sensing and ventricular pacing with heart rate 80 beat per minute (**Figure 9**).

## Discussions

Manipulation of a pacemaker or implantable cardioverter defibrillator that caused malfunction of the device has been known as Twiddler syndrome. The pacemaker is most often rotated on its long axis within the pocket.<sup>2</sup> Pacemaker rotation leading to lead displacement was previously described by Bayliss, et al. It was named after patients who twiddled with the device resulting in rotation and lead dislodgement. However, the syndrome can also occur spontaneously.<sup>3</sup>

For the most part, it is a painless phenomenon and the majority of patients do not claim a history of manipulating their device. It is more common in the elderly, presumably due to the laxity of their subcutaneous tissues.<sup>1,5</sup> Other risk factors include obesity, female, psychiatric illnesses, and size of the implanted device is relatively smaller than its pocket.<sup>3</sup> Manipulation may cause axial rotation of the pulse generator, twisting, and eventual fracture or dislodgment of the lead. The pulse generator is not usually damaged.<sup>4</sup> For obvious reasons, it can have dangerous consequences.<sup>6,7</sup>



**Figure 9.** ECG of post reposition lead and generator pacemaker showed atrial sensing ventricular pacing rhythm with 80 bpm heart rate.

The majority of cases occur during the first year of implantation, a “late Twiddler syndrome” has also been reported.<sup>16</sup> So far the earliest reported case is at 17 hours.<sup>1</sup> Our patients were detected to have Twiddler syndrome within eight and second months of their respective implantation. Twiddler syndrome of ICDs and pacemaker is typically attributed to neither conscious nor unconscious rotation of the generator. However, sometimes there is lack of evidence of twisting of the generator. The risk factors in these patients include elderly with weak subcutaneous tissue and subcutaneous pocket. Both patients were already retired and have history of driving motorcycle regularly as the most excessive physical exercise that pose as risk of this unusual complication. The movement of upper arm permitted dislodgement of the inserted unit. The increased muscular activity of the shoulder and arm might have led to repetitive motions within the pocket favoring the coiling of the leads.<sup>8</sup> In recent report, hard physical training during neurological rehabilitation has been associated with the twiddling of the proximal trans venous defibrillation lead.<sup>9</sup> During abduction of the arm, the generator moved cranial, pushing the leads upward that may then be twisted to form a loop. If a cogwheel phenomenon is present, the loop does not unwind when the arm is brought down again. When the movement is repeated, further twirling occurs.<sup>10</sup>

It may also explain the reported recurrence of the syndrome after revision of the system despite precautions like anchoring the generator to the fascia.

It seems prudent not to leave redundant lead between the site of insertion and the generator pocket to prevent the formation of a loop of the lead. Regular evaluation every 6 month of the position of ICD and PPM leads with fluoroscopy or chest X-ray is advisable in patients who take up more vigorous exercise involving extensive abduction of the arm.<sup>10,11</sup>

The lead dislodges, resulting in pacemaker malfunction, non-capture, and unintended stimulation of nearby nerves. It causes failure to pace and can result in stimulation of the brachial plexus, vagus nerve, pectoral muscle, or the phrenic nerve.<sup>3</sup> Signs and symptoms include abdominal pulsations, diaphragmatic pacing, presyncope, and syncope. The left brachial plexus may be stimulated by the dislodged lead, as in this first case, causing rhythmical twitching of the left pectoral muscles and abdominal pulsations.<sup>2</sup> No evidence of stimulation of nerves in the second case was found, but there were symptoms of dyspnea on effort similar before he underwent pacemaker implantation.

Standardized device implantation including fixation of proximal leads with non-absorbable double sutures around the sleeves to the pectorals muscle should be done to prevent migration. Commonly the device is placed in the upper pectorals muscle but sometimes it needs to be placed subpectoral or inframammary. When the subpectoral is needed, the greater pectoral muscle is divided between its subclavian and thoracic part in order to form a pocket beyond the pectorals muscle. After the hemostasis, fluoroscopy is advisable before the wound is closed

to ensure leads position. During hospitalization, restriction of patient's arm movement is advisable. Preventive measures include using a compression band around the upper chest and shoulder, and tightening of the patient's arm for at least five to seven days. An anteroposterior and lateral position of chest X-ray should be made to show leads placement and rule out pneumothorax.<sup>11</sup>

Twiddler syndrome is more frequent in older female and obese patients probably due to more subcutaneous space between cutis and pectorals muscle. Fixating device at pectoral fascia and pectoral muscle may prevent this complication.<sup>3,14</sup> Fixating of the device-header with one suture would probably have not prevented the generator from being rotated, so anchoring the device into tightly fitting subpectoral pocket will prevent twiddling the device around that suture.<sup>12</sup> Management options for preventing or treating twiddling may include positioning the pacemaker in an abdominal pocket because there was no increased mobility of the device in an abdominal pocket. The reduction of the pocket size by creating a smaller pacemaker pocket to fit the smaller device can prevent the development of the late version of the Twiddler syndrome. The miniature size of the new devices permits their rotation during physical activities when positioned in the old pacemaker's pocket.<sup>13,14,15</sup> Preventive measures such as patient education will reduce the risk of developing the syndrome. Individualized patient education is important.<sup>2</sup> A lot of patients who need implantation of an ICD or pacemakers are in older age. Therefore it is important to suggest not only the patient but also their family to avoiding an overuse of upper arm that ipsilateral to the site of device implantation.

## Conclusion

Lead dislodgement in Twiddler syndrome is potentially dangerous especially in pacemaker dependent patient. In patient with implantable cardioverter-defibrillators (ICDs), it may not manifest as cardiac symptom but can result in stimulation of brachial plexus, vagus nerve, pectoral muscle, or phrenic nerve causing rhythmical twitching of left pectoral muscles and abdominal pulsations. Important management options for preventing or treating twiddling may include fixating the device at pectoral fascia, anchoring the device into tightly fitting subpectoral

pocket, creating a smaller pacemaker pocket and adequate individualized education for patient and family. Regular evaluation every 6 month of the position of ICD and PPM leads with fluoroscopy or chest X-ray is advisable.

## Abbreviations

bpm: beat per minute

DCM: dilated cardiomyopathy/*kardiomiopati dilatasi*

ECG: electrocardiography

ICD: implanted cardioverter-defibrillator

PPM: permanent pacemaker

RV: right ventricle

RVO: right ventricular outflow tract

TAVB: total atrioventricular block/*penyumbatan atrioventrikular total*

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