



## Twiddler's Syndrome: A Rare but Lethal Complication

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### Introduction

Twiddler's syndrome is a rare but potentially fatal cause of pacemaker or implantable cardioverter defibrillator (ICD) malfunction. It happens due to the patient's unintentional or intentional manipulation of the generator, causing painless torsion and leads dislodgment that results in permanent loss of device function<sup>1,2,3</sup>.

### Case Report

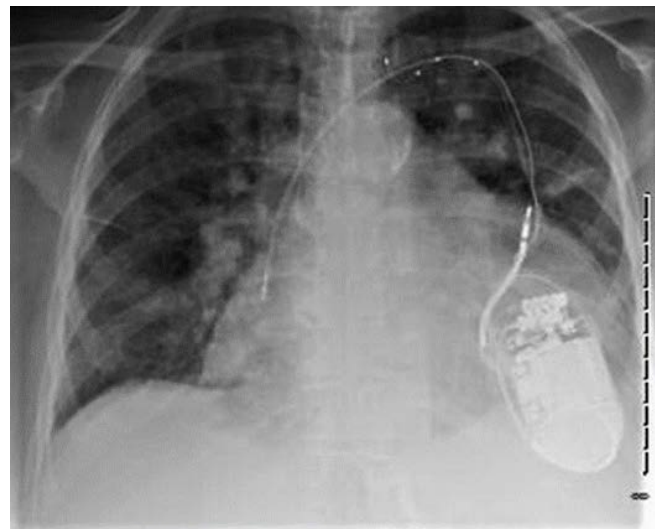
A 68-year-old female with a history of nonischemic dilated cardiomyopathy, class III congestive cardiac failure, and bifascicular block presented to the outpatient clinic with sudden worsening of bilateral ankle edema.

She underwent a biventricular pacemaker/defibrillator placement 3 months prior for primary prevention of sudden cardiac death. A follow-up chest X-ray 1 week after the biventricular pacemaker/defibrillator placement was unremarkable (figure 1).



**Figure 1:** Chest X-ray showing proper placement of implanted leads and pacemaker/defibrillator device 1 week after implantation.

On this arrival, an electrocardiogram (EKG) showed pacing spike artifacts without appropriate capture. A chest X-ray revealed that the atrial and ventricular leads were out of the vascular space and wrapped around the pacemaker in the pacemaker pocket (figure 2). Interrogation of the device showed complete loss of pacing and sensing functions.



**Figure 2:** Chest X-ray showing implanted leads dislodgement with pacemaker/defibrillator device displacement into lower chest due to manipulation by the patient.

Upon questioning about pacemaker manipulation, the patient admitted twisting her pacemaker generator frequently. She was immediately admitted to the hospital for repositioning of the leads and pacemaker generator. All leads were repositioned properly except the coronary sinus lead. The device was repositioned in the pocket and sutured to the underlying pectoralis muscle to prevent recurrent manipulation. Follow-up chest X-ray and EKG were unremarkable. The patient and her caregiver were educated

that the most important step to prevent recurrence is to avoid twiddling the pacemaker generator. Ongoing follow-up visits were unremarkable.

## Discussion

Twiddler's syndrome is pacemaker malfunction due to the patient's repeated twisting of the pacemaker pulse generator, which causes painless leads dislodgement and device rotation.

This syndrome is common in patients with compulsive disorders<sup>1,4</sup>. Elderly and obese females are also at high risk due to increased laxity of subcutaneous tissues that allows for easier rotation of the pulse generator in its pocket.

Most reported cases happened within the first year of implantation<sup>1,3</sup>. However, Twiddler's syndrome can occur anytime after the implantation<sup>1</sup>. The earliest reported case happened 17 hours after pacemaker placement<sup>3</sup>.

A dislodged lead can stimulate the ipsilateral phrenic nerve resulting in abdominal pulsation or hiccups. Coiled leads can stimulate brachial plexus causing rhythmic arm twitching<sup>2</sup>. Twiddler's syndrome could be fatal in a patient with a defibrillator due to uncomfortable, inappropriate shocks, and inability to deliver shocks when needed in true malignant ventricular arrhythmias because of loss of adequate sensing and capture<sup>4</sup>.

Fast diagnosis and management are crucial. EKG, chest X-ray, and pacemaker interrogation can give the essential information to diagnose Twiddler's syndrome. The chest X-ray is vital for visualizing the device and the implanted lead displacement, both of which are needed to diagnose Twiddler's syndrome<sup>2,3</sup>.

Treatment of Twiddler's syndrome includes readmission, lead uncoiling, new lead implantation, repositioning of the dislodged leads, and pulse generator fixation<sup>2</sup>. Appropriate implantation technique is one of the solutions to prevent Twiddler's syndrome that includes creating a small surgical pocket and subpectoral device placement<sup>3,4</sup>. Patients with mental disorders and cognitive dysfunction who have proven Twiddler's syndrome may benefit from the recently

approved leadless MICRA transcatheter pacing system (TPS) when appropriate<sup>5</sup>.

Patient and family education against further device manipulation is critical for the long-term prevention of Twiddler's syndrome<sup>4</sup>. Physicians should be vigilant about the high-risk patients who develop Twiddler's syndrome: elderly, obese females, patients with mental disorders, and patients with cognitive dysfunction to ensure timely detection and management<sup>3,4</sup>.

## Conclusion

- Twiddler's syndrome should always be in the differentials as a cause of pacemaker failure in the elderly patient presenting with bradyarrhythmias, syncopal episodes after pacemaker implantation<sup>1,3</sup>.
- The chest X-ray and EKG are the vital diagnostic tests for Twiddler's syndrome<sup>1,3,4</sup>.
- Patient and caregiver counseling against manipulating the pacemaker generator is paramount to prevent this potentially life-threatening complication<sup>3,4</sup>.

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## Conflicts of interest

There are no conflicts of interest.

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