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CASE REPORT
Arrhythmias/electrophysiology

Twiddler's syndrome with a subcutaneous implantable cardioverter-defibrillator presenting with an inappropriate shock: a case report

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Background Subcutaneous implantable cardioverter-defibrillators (S-ICDs) are increasingly used in patients at risk of fatal cardiac arrhythmias. Twiddler's syndrome is a condition in which a device is manipulated by the patient after implantation leading to lead twisting and retraction. Device manipulation has been reported multiple times in transvenous pacing systems and occasionally leads to inappropriate discharges from implanted defibrillators. However, little has been reported about device manipulation in S-ICD devices.

Case summary We present the case of a 16-year-old who underwent insertion of an S-ICD for idiopathic dilated cardiomyopathy. He represented for a pacing check following a discharge from the device. This showed a significant change in the sensed vectors. Chest radiographs confirmed lead retraction and suggested device manipulation. The device was turned off to prevent further inappropriate shocks. The patient underwent successful reimplantation of a S-ICD device.

Discussion This case highlights that twiddler's syndrome can occur in those with an S-ICD and lead to an inappropriate device discharge. The patient in this case had a number of risk factors that have been previously associated with twiddler's syndrome.

Keywords Case report • Twiddler's syndrome • Subcutaneous ICD

Learning points

- Twiddler's syndrome should be considered prior to implantation in subcutaneous implantable cardioverter-defibrillator (S-ICD) cases as well as transvenous cases.
- Twiddler's syndrome can lead to a significant change in sensed vectors and result in inappropriate device discharge in S-ICDs.

Introduction

Twiddler's syndrome is an uncommon condition in which an implanted device, a pacemaker or implantable cardioverter-defibrillator (ICD), is manipulated by the patient, resulting in lead displacement. This has been recognized in transvenous pacemakers, ICDs, deep brain stimulation devices,[1] and implanted pump devices.[2] This

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condition was first recognized by Bayliss *et al*. in 1968.[3] Lead macrodisplacement (LMD) has been reported as an uncommon complication in patients with transvenous pacemakers and ICDs.[4,5] In the case of pacemakers, the condition often presents with failure of the device; in ICDs it can lead to inappropriate shocks.[4]

Subcutaneous ICDs (S-ICDs) are composed of a subcutaneous electrode placed along the sternum and a generator placed below the left axilla.[6] Patient selection is important prior to S-ICD implantation to prevent inappropriate shocks through T-wave oversensing.[7] It has been suggested that the use of S-ICDs may reduce systemic infections and other complications that occur with repeated replacement of transvenous ICDs and specifically avoid the complications related to venous access and having leads in the vascular system; this is currently being investigated in a randomized controlled trial.[8] We present a case of twiddler's syndrome occurring in a patient with a S-ICD, presenting with an inappropriate shock.

Timeline

Date	Events
Day 0	The patient was admitted to local hospital with signs and symptoms consistent with congestive cardiac failure
Day 1	The patient was transferred to a specialist paediatric cardiology centre where he underwent a transthoracic echocardiogram showing a left ventricular ejection fraction (LVEF) 10%
	The patient was diagnosed with dilated cardiomyopathy and an associated left ventricular apical thrombus
Day 151	Follow-up transthoracic echocardiogram was performed showing an LVEF 10% and no evidence of thrombus
Day 166	A decision was made with the patient to implant an subcutaneous implantable cardioverter-defibrillator (S-ICD)
	Referral made to advanced heart failure service for monitoring and consideration of heart transplantation if the patient deteriorated
	Referral to genetic screening service made
Day 199	Insertion of a primary prevention Boston Scientific S-ICD under general anaesthetic
12 months	Routine review in heart function clinic
	No implantable cardioverter-defibrillator (ICD) therapies had occurred during this time
18 months	Attended pacing clinic for review following inappropriate ICD shock
	Retraction of subcutaneous lead noted on chest radiograph and change in sensing vectors apparent on interrogation of the device
21 months	Reimplantation of an S-ICD under general anaesthetic

Case presentation

The male patient initially presented to clinical care at the age of 16 years with increasing shortness of breath, peripheral oedema, and orthopnoea. Past medical history included pathological hyperphagia and obesity. Clinical examination on initial presentation revealed body mass index (BMI) of 33.4 kg/m^2, bi-basal crackles on respiratory auscultation, and pitting oedema to the knees. Cardiac auscultation demonstrated normal heart sounds without any murmurs. During his initial admission, he was transferred to a specialist paediatric cardiology centre for ongoing care. An echocardiogram demonstrated a dilated left ventricle with an ejection fraction (LVEF) 10% and a left ventricular thrombus. He was initiated on warfarin (target INR 2–3), bisoprolol 2.5 mg o.d., ramipril 2.5 mg b.i.d., and spironolactone 25 mg o.d. He underwent psychiatric assessment whilst an inpatient and was commenced on sertraline 100 mg, principally to assist with appetite control.

Following discharge, the patient was reviewed in the local heart function clinic. A repeat echocardiogram confirmed an LVEF of 10% and an electrocardiogram showed a narrow QRS complex. A decision was made with the patient and his mother for him to have a S-ICD fitted for primary prevention.

The patient underwent screening investigations to ensure he was suitable for S-ICD implantation. The implantation was performed under general anaesthetic and was uncomplicated. The S-ICD was implanted using an intermuscular approach with the device placed between the anterior surface of serratus anterior and the posterior surface of latissimus dorsi.[9] A ventricular fibrillation induction test was performed at implant, with successful cardioversion. A chest radiograph confirmed appropriate positioning of the lead and generator (*Figure 1*) and appropriate sensing vectors were confirmed post-implant (*Figure 2*).

He was reviewed 6 months later and reported a general improvement in symptoms. No therapies were recorded from the device in this timeframe. Thirteen months after device implant (October 2018), the patient presented emergently to the pacing clinic, having experienced a shock. On interrogation, it was noted that the vectors on the device had changed significantly (*Figure 3*); this prompted a repeat chest radiograph (*Figure 4*). It was noted that there had been retraction of the subcutaneous lead and coiling of the lead around the generator. Given the presentation with an inappropriate shock, the device was switched off.

On direct questioning, the patient stated that he was not aware of any conscious manipulation of the device. It was agreed that repositioning of the S-ICD lead was in his best interest, and this was undertaken successfully in December 2018 with the device again placed intermuscularly between serratus anterior and latissimus dorsi. Routine device follow-up in February 2019 has revealed normal sensing vectors (*Figure 5*) and no further shocks.

Discussion

This case highlights the risk of twiddler's syndrome in the S-ICD population and the potential for this to result in inappropriate therapies. In a review of the literature, only one other case of twiddler's syndrome in association with an S-ICD has been reported. Unlike the

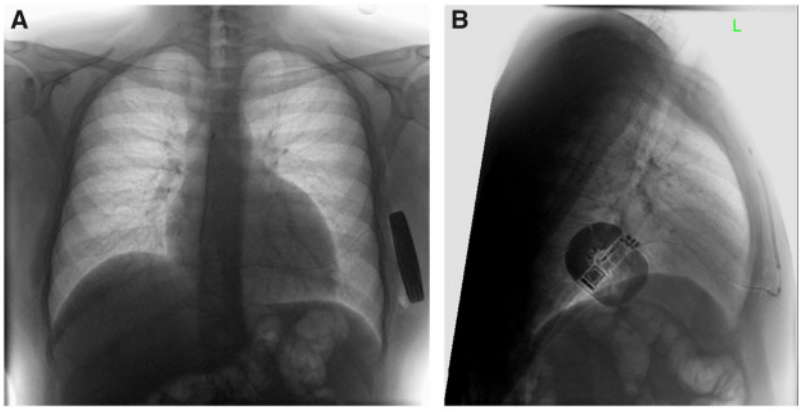

Figure 1 Chest radiograph following device implantation showing correct lead position. (*A*) PA projection. (*B*) Lateral projection.

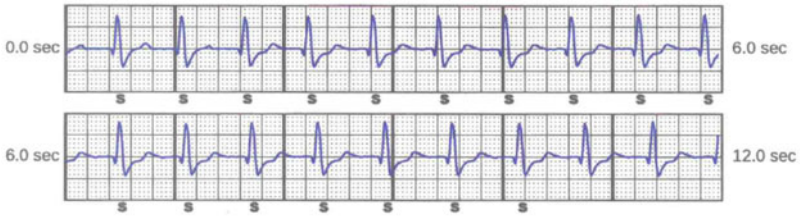

Figure 2 Sensing vectors after initial implant.

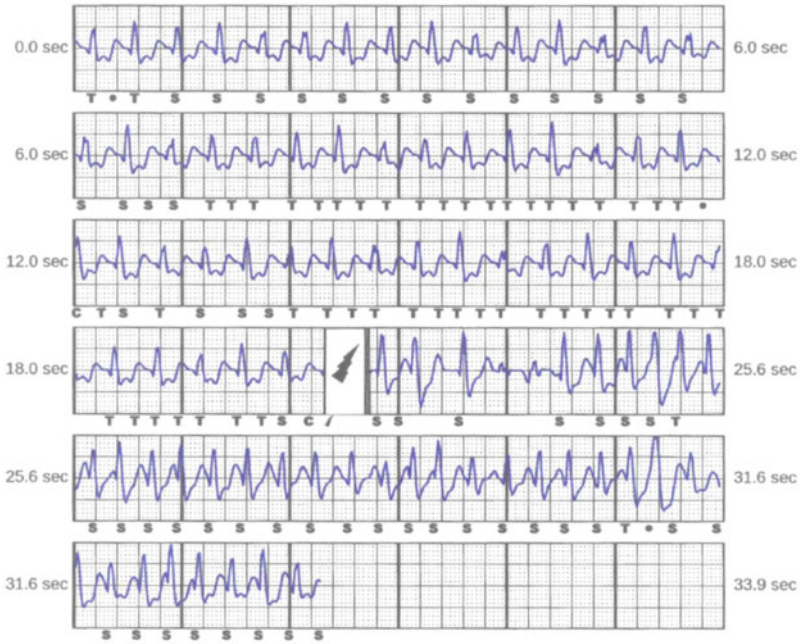

Figure 3 Sensing vectors after the patient presented for an inappropriate shock.

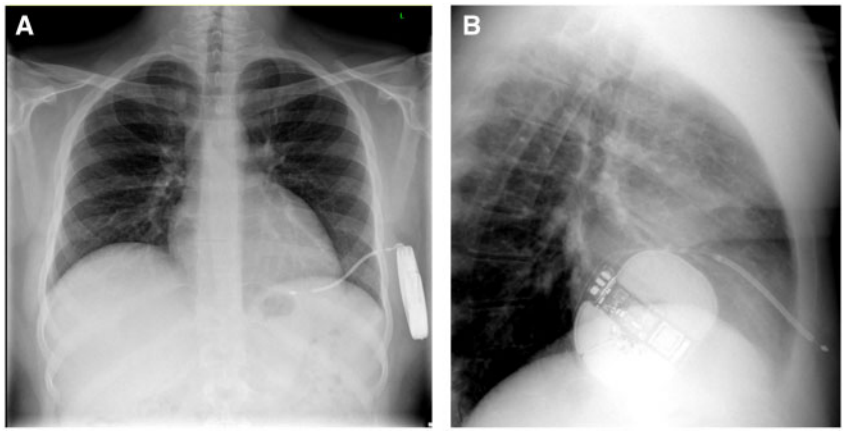

Figure 4 Chest radiograph after an inappropriate shock showing retraction of the chest lead back towards the subcutaneous implantable cardioverter-defibrillator generator. (*A*) PA projection. (*B*) Lateral projection.

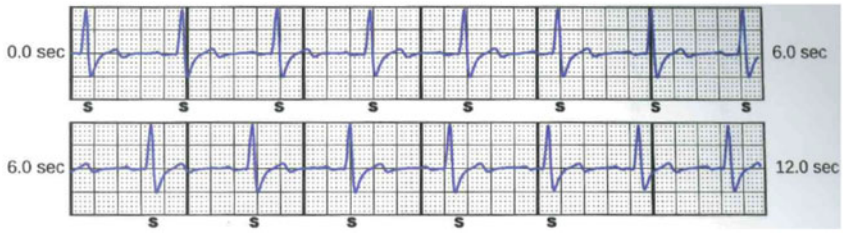

Figure 5 Sensing vectors after lead repositioning.

case described, that patient presented with chest pain and a loss of sensing from the device.[10]

The risk of inappropriate therapies has previously been recognized to be higher in patients with S-ICDs compared to those with transvenous ICDs; however, a recent meta-analysis has shown similar rates of total inappropriate therapy between S-ICD and transvenous systems.[11] The higher rate of inappropriate therapies previously seen in S-ICD patients is largely due to subcutaneous electrodes, rather than an intracardiac electrogram, interpreting the cardiac rhythm.[12] Prescreening prior to S-ICD insertion attempts to reduce the likelihood of a patient experiencing an inappropriate shock, particularly due to T-wave oversensing. Twiddler's syndrome causing lead retraction in patients with an S-ICD will significantly alter the sensed vectors, predisposing to inappropriate device discharge.

Twiddler's syndrome is a form of LMD. A recent retrospective cohort analysis of transvenous cardiac devices revealed that LMD occurred in 1.8% of cases (total cohort = 1074)[4]; however, only one case was reported as twiddler's syndrome. Of 19 cases with LMD, eight were ICDs, of these two presented with inappropriate shocks. Increasing pacing thresholds/lead impendence are commonly noted in twiddler's syndrome.[13] In this case, the dramatic alteration in sensing across all three vectors prompted radiographic re-evaluation of lead positioning, and established the diagnosis.

A number of risk factors have previously been described for the development of twiddler's syndrome. Those most commonly quoted include: female gender,[4,13] increased BMI,[4,13] paediatric patient,[14] elderly patient,[13] past mental health history,[14] and device-pocket size mismatch.[15] In retrospect, the patient described fulfils a number of these risk factors (increased BMI, paediatric patient, and history of psychiatric disorder). The S-ICD device and leads are secured with sutures, but recognition of the higher risk twiddler patient might necessitate extra sutures for example. Use of the three incision technique should be considered to reduce risk of lead dislocation.

Conclusion

Lead displacement due to manipulation of the generator in patients with S-ICD can result in inappropriate shocks. Possible risk factors for this include obesity and a history of psychiatric illness. Careful assessment of the balance in favour of S-ICD over transvenous ICD should be made in patients with these conditions.

Lead author biography

C. Fielder Camm is a cardiology registrar working in the Cardiology Department of the Royal Berkshire Hospital. He has a clinical interest in cardiac devices and electrophysiology. He is currently the Assistant Editor of *European Heart Journal - Case Reports*.

Supplementary material

Supplementary material is available at *European Heart Journal - Case Reports* online.

Slide sets: A fully edited slide set detailing this case and suitable for local presentation is available online as Supplementary data.

Consent: The author/s confirm that written consent for submission and publication of this case report including image(s) and associated text has been obtained from the patient in line with COPE guidance.

Conflict of interest: C.F.C. is the Assistant Editor of *European Heart Journal - Case Reports*.

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