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CASE REPORT
Arrhythmias/electrophysiology

Ratchet mechanism selectively causing idiopathic macrodislodgement of an active-fixation coronary sinus lead: a case report

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Background ‘Idiopathic’ lead macrodislodgement may be due to Twiddler’s syndrome depending on active twisting of pulse generator within subcutaneous pocket. All leads are involved, at any time from implantation, and frequently damaged. In the past few years, a reel syndrome was also observed: retraction of pacemaker leads into pocket without patient manipulation, owing to lead circling the generator. In other cases, a ‘ratchet’ mechanism has been postulated. Reel and ratchet mechanisms require loose anchoring, occur generally briefly after implantation, with non-damaged leads. We report the first case of an active-fixation coronary sinus lead selective macrodislodgement involving such ratchet mechanism.

Case summary A 65-year-old man underwent biventricular defibrillator device implantation, with active-fixation coronary sinus lead. Eight months later, he complained of muscle contractions over device pocket. At fluoroscopy, coronary sinus lead was found near to pocket, outside of thoracic inlet. Atrial and ventricular leads were in normal position. After opening pocket, a short tract of coronary sinus lead appeared anteriorly dislocated to generator, while greater length of lead body twisted a reel behind. The distal part of lead was found outside venous entry at careful dissection. Atrial and ventricular leads were firmly anchored.

Discussion Our case is a selective ‘Idiopathic’ lead macrodislodgement, possibly due to a ratchet mechanism between the lead and the suture sleeve, induced by normal arm motion; such mechanism incredibly, and for first time in literature involves a coronary sinus active-fixation lead.

Conclusion Careful attention should always be paid to secure anchoring even of active-fixation coronary sinus leads.

Keywords Reel syndrome • Twiddler’s syndrome • Ratchet mechanism • Coronary sinus lead dislodgement • Coronary sinus active-fixation lead • Case report

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Learning points

- Coronary sinus (CS) lead dislodgement can occur after implantation, and stability issues due to CS venous anatomy play the main role; consequently, active-fixation CS leads have been released, with good results.
- Twiddler's and reel syndromes are rare but well-known mechanisms responsible for 'Idiopathic' lead macrodislodgement (ILMD); some ratchet mechanism between the lead and the suture sleeve can also cause ILMD.
- We report the first case of an active-fixation CS lead selective ILMD involving such a ratchet mechanism.
- Careful attention should be paid to secure tightening the suture sleeve, firm anchoring the pulse generator, adequate tailoring and sizing of the pocket to the dimensions and shape of the device can. The rotating movement used to deploy and fix the anchoring helixes and the smaller size of the active-fixation CS lead could have played a role in this specific case.

Introduction

Twiddler's syndrome consists of twisting of the pulse generator (PG) within the subcutaneous pocket, sometimes by active manipulation by the patient, resulting in dislodgement and retraction of the pacing leads that usually are simultaneously involved as a whole. In the past few years, another type of 'idiopathic"' lead macrodislodgement (ILMD) was observed, the reel syndrome: a 'spontaneous' retraction of the pacemaker (PM) leads into the pocket without patient manipulation, with the leads lying retracted and encircling the generator, in the absence of twisting.

In very few cases, only a single lead retracts into the pocket while the other leads are not involved, and the consistent interpretation is not yet clear. We describe the first case of an ILMD syndrome selectively involving an active-fixation coronary sinus (CS) lead as part of a biventricular implantable defibrillator device without affecting the two other leads.

Timeline

Case	A 65-year-old male caucasian patient, with coronary artery disease, hypertension, diabetes mellitus, obesity (BMI 32); with Class III NYHA symptoms despite optimal medical therapy, complete left bundle branch block, severely depressed ejection fraction, about 25–30%.
18 October 2018	CRT-D implantation.
21 October 2018	Normal postoperative chest X-ray, normal electrical parameters at hospital discharge.
20 November 2018	Normal 1-month follow-up.
Late December 2018	Muscle contractions over right haemidiaphragm
27 December 2018	Left ventricular (LV) stimulation turned off. No chest X-ray repeated, nor standard 12-lead electrocardiogram registered.
7 April 2019	While performing threshold testing, muscle contractions observed over the device pocket, with standard bipolar pacing. Failure to pace even at maximal output, no sensing. Chest X-ray performed, showing the dislodgment of the LV lead.
14 June 2019	Implant revision. Left ventricular lead tip found outside the venous entry, probably due to ratchet mechanism. Other leads in stable position. Coronary sinus (CS) lead extracted; new CS lead implanted.
17 June 2019	Stable CS lead position; normal electrical function; healthy healing surgical wound.
15 July 2019; 24 September 2019; 20 January 2020, June 2020	No recurrence of muscle contractions. Normal ambulatory device checks, with concomitant improvement of ejection fraction and functional class

Continued

Case presentation

A 65-year-old male caucasian patient with a history of ischaemic heart disease, hypertension, diabetes mellitus, obesity (BMI 32), presented with progressive congestive heart failure, unresponsive to optimal medical therapy, and complete chronic left bundle branch block (LBBB). He complained of Class III NYHA symptoms with a severely depressed ejection fraction, about 25%. This patient underwent a CRT-D device implantation on 18 October 2018 (Amplia MRI Quad CRT-D, Medtronic). The right atrial (RA) and the right ventricular (RV) leads were inserted through surgical cut-down of the cephalic vein, and the CS left ventricular (LV) lead through intrathoracic left subclavian vein puncture. An atrial J tined lead (Capsure Sense 4574, Medtronic), a single-coil active-fixation shock lead (Sprint Quattro Secure S 6935M), and an active-fixation LV lead (Attain Stability Quad, Mod. 4798, Medtronic) were used. All leads were firmly anchored through suture sleeves to the pectoral fascia. The generator was located in a subcutaneous pocket, and not sutured to the fascia, as usual. The leads appeared in the normal position on the postoperative chest X-ray, with the normal electric behaviour during the device interrogation. In late December 2018, the patient complained of right diaphragmatic contractions. During a fast PM check in another

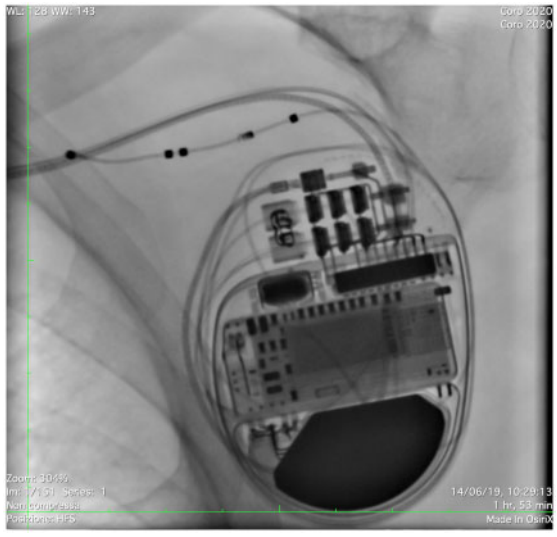


Figure 1 Fluoroscopy showing the left ventricular lead reeled into the pacemaker pocket, encircling the pulse generator with its tip retracted in close proximity of the generator can.

Centre, LV stimulation was turned off. At that moment, a 12-lead standard electrocardiogram was not recorded; moreover, a chest X-ray was not repeated, due to lack of time, being the patient abroad for Christmas holidays and hurrying home. However, a quick further and closer examination at home was recommended. Even though if the patient was suffering from low-level exertional dyspnoea (NYHA Class III), due to absence of the irritating muscle contractions, the PM interrogation was only performed, as scheduled, 6 months after the implantation, on April 2019. Threshold testing showed loss of capture of the LV lead, even at maximal energy. On the contrary, the test repeatedly triggered muscle twitching on the left upper part of chest, mainly in close proximity of the CRT-D pocket; such a behaviour was clearly observed also with the standard bipolar pacing configuration, even with the low-energy outputs. The RA and RV lead pacing and sensing parameters were unchanged. A postero-anterior chest X-ray disclosed individual displacement of the CS lead. Fluoroscopy in multiple views showed the LV lead reeled into the PM pocket, encircling the PG with its tip retracted in close proximity of the PG can (*Figure 1*). The RA and RV leads maintained their correct position, without any change. No displacement of the PG was detected.

No apparent cause for reeling of the lead was evident at history. The patient strictly denied any history of manipulation or trauma over the pocket area or the PG. Significant hyperabduction of the left arm or traction along the lead was excluded.

The patient was admitted to hospital on June 2019.

Physical examination showed a heart rate of 90 b.p.m., blood pressure of 95/60 mmHg, with a S3–S4 gallop, a holosystolic apical murmur with radiation to left axilla and a proto-mesosystolic murmur in the subxiphoid area. There was jugular venous distension with an enlarged tender liver, and ankle and pretibial oedema. Signs of peripheral hypoperfusion and right-heart failure were present (vasoconstriction, cyanosis, jaundice), with laboratory examinations consistent with hepatic and renal involvement (AST 50 U/L, ALT 45 U/L, and serum creatinine 2.9 mg/dL). Electrocardiogram showed a complete LBBB with left-axis deviation. Two-dimensional echocardiogram showed severe dilatation of the left ventricle (LVEDD 66 mm) due to diffuse hypokinesia, with depressed ejection fraction (EF) (28%); moderate-to-severe functional mitral regurgitation and moderate tricuspid regurgitation, with pulmonary hypertension (PAPs 60 mmHg).

Surgical revision was performed on 14 June 2019. Careful dissection showed the LV lead rolling around and before the PG, like a fishing line around a reel (*Figure 2A*; *Video 1*). The LV lead was widely movable, in part free into the pocket, in part adhering to the pocket capsula (*Figure 2B* and *C*; *Video 2*). At the end of dissection, without performing any traction, the LV tip was found outside the entry point of left subclavian vein, in close proximity of the PG (*Figure 2D*; *Video 3*). On the contrary, the atrial and shock leads appeared firmly secured in the correct position and wrapped into the capsular fibrous coating layer (*Figure 2B* and *C*; *Video 2*).

The LV lead was then extracted, and a new LV passive-fixation lead was implanted in the same previously used postero-lateral vein. One day after the procedure chest X-ray showed all leads correctly placed, with normal electric function, and with a normal healing surgical wound. The subsequent ambulatory device checks were normal, after 1-year follow-up, with concomitant improvement of EF (38%) and functional class (NYHA Class II).

Discussion

Twiddler's syndrome was first described by Bayliss in 1968,[1] as a cause of ILMD. It is known to occur more often in elderly women or in overweight patients with increased laxity of the subcutaneous tissue, with large pockets, as well as in patients with psychiatric disorders. The mechanism of twiddler's syndrome is twisting the generator unconsciously or more often consciously by the patient.

The term 'reel syndrome', instead, was first coined by Carnero-Varo *et al.*[2] in 1999 to describe a specific form of twiddler's syndrome in a patient with a single-chamber PM. Although the mechanism was not clearly determined, it may have been due to rotation of the PG on its transverse axis resulting in reeling in of the PM lead around to the can, like a fishing reel. This spontaneous retraction of PM leads into the pocket may occur without patient manipulation, sometimes due to a large subcutaneous pocket. There's a basic difference between twiddler's and reel syndromes in the former, the device rotates along its longitudinal axis, while in the latter it rotates along the transverse axis.[3] Both syndromes usually involve all implanted leads, especially twiddler's syndrome. Uncommonly, one lead only is implicated, and retracts and dislocates,[3] while the other leads maintain their normal position. In 2007, Von Bergen *et al.*[4] speculated a 'ratchet' mechanism involving the lead and the suture sleeve as the possible explanation.

Twiddler's and reel syndromes are commonly due to an excessive PG movement in a relatively oversized pocket. Generally, reel and ratchet syndromes require sufficiently loosened suture ties to permit the retraction of the lead; they occur almost briefly after the implantation, without a direct damage of the lead.[5] On the contrary, the

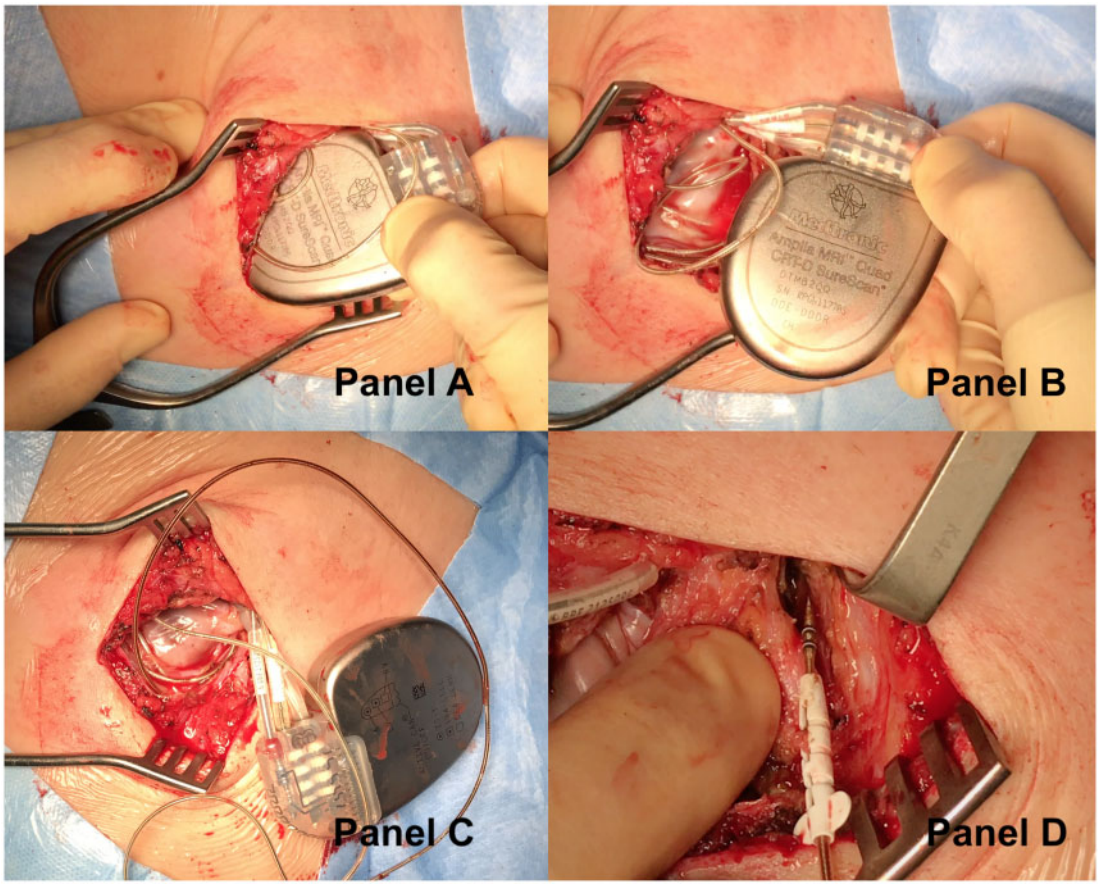
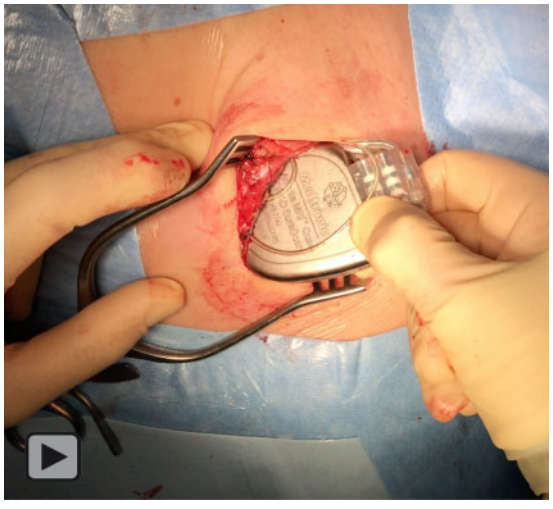
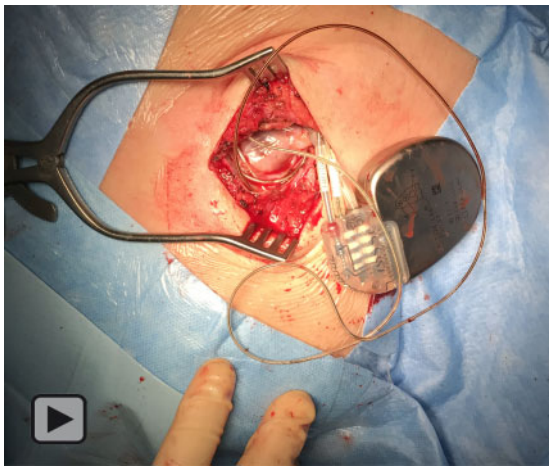


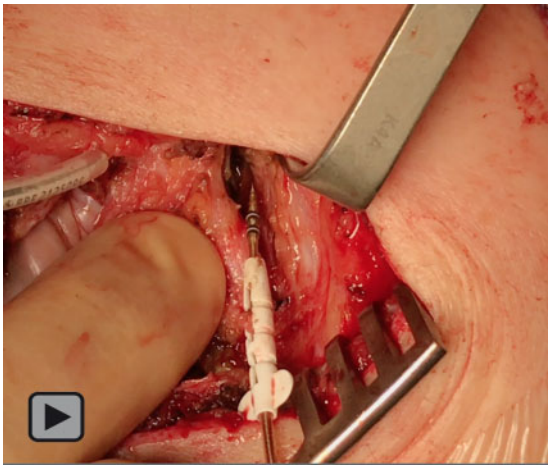
Figure 2 Dissection along the left ventricular lead. (*A*) Pocked opening. The left ventricular lead is rolling around and before the pulse generator. (*B* and *C*) Wider careful dissection, showing the left ventricular lead widely movable, in part free into the pocket, in part adhering to the pocket capsula. Note the atrial and shock lead firmly secured in the correct position and wrapped into the capsular fibrous coating layer. (*D*) At the end of dissection, without performing any traction, the left ventricular tip is found outside the venous entry.



Video 1 Pocket opening. The left ventricular lead appears anteriorly located to the pulse generator, and encircling it.



Video 2 Dissection/mobilization of left ventricular lead. A wide careful dissection is performed, showing the left ventricular lead extensively mobilizable, in part free into the pocket, in part adhering to the pocket capsula. The atrial and shock lead appear firmly secured in the correct position and wrapped into the capsular fibrous coating.



Video 3 Left ventricular tip outside venous entry. End of dissection. Without performing any traction, the left ventricular tip is found outside the venous entry.

twiddler's syndrome can take place at any time after PM implantation, and the leads are often damaged. Specifically, the ratchet syndrome is due to the unidirectional withdrawal of the lead. Typical risk factors for the ILMD, like female sex, obesity, concomitant psychiatric disorders have not been clearly associated to these syndromes in the recent years.[6]

Some procedural factors, like a lateral incision, the proximity of shoulder joint, multiple leads, passive-fixation leads, loose suturing of fixation sleeves, the device/pocket mismatch and the incomplete/absent securing the device into the pocket have been postulated as the predisposing mechanisms.[6] An increased laxity and thickness of the subcutaneous tissue may also contribute. Direct trauma and repetitive shoulder movements have been recognized as the immediate efficient causes.

By examining the first postero-anterior X-ray and after a careful fluoroscopy in multiple views, we hypothesized a ratchet mechanism selectively involving the CS lead, between the lead and the suture sleeve. This mechanism, rather than the twiddler's or reel syndromes, seemed more consistent with the intra-operative examination, disclosing an unchanged position of the device case, so excluding a PG rotation on one of its axes.

The ILMD syndromes have been described also for defibrillators a lot of years ago,[7] owing to lead failure,[8] and also to multiple inappropriate shocks.[9,10] Also CRT-D devices can be involved,[11,12] even causing worsening of the resynchronization efficacy.[13] The RV active-fixation leads may be extraordinarily affected.[14]

Like in the paper of Von Bergen *et al.*,[4] indeed, in our case only the LV lead has been involved. It can be assumed that a slight tug on the lead pulls it and a ratchet mechanism involves the lead through the suture sleeve, enabling a one-way movement towards the PG, without being able to drive it back to its original position. We further hypothesize that a ratchet selective syndrome can occur with a normal arm motion.

Isolated ratchet dislodgement like our one is rare in literature. Spontaneous retractions of LV leads by ratchet mechanism have been described.[15] In all these two cases, ILMD was observed only with passive-fixation LV leads, and always post a CRT-D generator change following extensive capsulectomy and dissection of the fibrous tissue around the suture sleeve and of the adherences along the leads.

However, no previous reported case involves an LV lead with an active-fixation mechanism. In this specific case, the Medtronic Stability Quad 4798 has active-fixation side helixes that are deployed by means of a rotational movement transmitted from the pin through the lead body up to the lead tip. We can suppose this counter-rotation of the lead may have facilitated the ratchet mechanism, through a rotational elastic recoil and back gliding inside a partially loosened fixation sleeve. Therefore, particular attention should be given to the fixation of all leads, especially in overweight patients with laxity of the subcutaneous tissue, with close ties on suture sleeves; in avoiding a great device/pocket mismatch and a very lateral incision near the shoulder joint; and in firmly securing to the fascia the device case, particularly in patient at risk.

Conclusion

Our case highlights the eventual occurrence of a selective reel syndrome not caused by the PG rotation, but possibly due to a ratchet mechanism between the lead and the suture sleeve, induced by normal arm motion, even in the absence of patient manipulation. Our case is the first describing this phenomenon involving an active-fixation CS lead, probably with a facilitating role of the rotational movement for fixing the lead, acting together with a partially loose fixation sleeve.

Limitations

The ratchet mechanism is one proposed hypothesis, but obviously cannot be directly proven. Such mechanism is strongly consistent with the finding of the LV suture sleeve in an unchanged position; the distal tip of the thin LV lead widely retracted outside the venous entry in close proximity of the PG can; and the other two leads not involved.

Lead author biography

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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 authors 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.

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