

Reel syndrome, a diagnostic conundrum: a case report

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Background

Pacemaker lead dislodgement and failure, related to device manipulation, is a rare complication of permanent pacemaker (PPM) insertion. Reel's, Twiddler's, and Ratchet syndrome are rare causes of pacemaker failure with varying mechanisms, defined by their classical lead and generator findings on chest X-ray imaging. Misleading patient presentations may be attributed to lead stimulation of surrounding structures.

Case summary

A 77-year-old female was admitted with abdominal wall pulsations, abdominal pain, and lower limb jerking 3 months following PPM insertion. Following exclusion of a ruptured abdominal aortic aneurysm, the presence of Reel syndrome was noted on the patient's chest X-ray and the electrocardiogram showed inappropriate pacing. Deactivation of the pacemaker resulted in immediate symptom cessation and urgent repositioning of pacemaker leads was undertaken.

Discussion

This case highlights the importance of considering pacemaker complications causing non-cardiac symptomatology. Pacemaker lead stimulation of surrounding structures can present in an unconventional fashion, veiling the diagnosis. However, a structured approach to undifferentiated neuromuscular presentations in patients with PPMs should consider lead dislodgement as a differential diagnosis. Rapid recognition of lead dislodgement, device deactivation, and re-implantation or repositioning of the leads are critical in preventing potentially life-threatening complications.

Keywords

Cardiac pacemaker • Lead dislodgement • Reel syndrome • Case report

Learning points

- Pacemaker lead dislodgement syndromes are rare and can present a diagnostic dilemma for clinicians. Timely identification and resolution of the issue can be critical in preventing life-threatening complications.
- The Reel syndrome is characterized by the 'reeling' of pacemaker leads around the generator and is caused by intentional or unintentional rotation of pacemaker generator along the frontal plane. It may present late (>6 weeks) after pacemaker insertion.
- Pacemaker lead dislodgement education to the patient and family members would play an important role in risk mitigation. Specific procedural techniques employed during pacemaker insertion may reduce the likelihood of device manipulation-related pacemaker lead dislodgement.

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Introduction

Pacemaker lead dislodgement and failure, related to device manipulation, is a rare complication of permanent pacemaker (PPM) insertion. It is reported to occur in up to 1.7% of implants, particularly in patients with large generator pockets or cognitive impairment.¹ Late (>6 weeks) complications of lead or electrode dislodgement are even more infrequent (~0.2%).² Reel, Twiddler, and Ratchet syndrome are rare causes of pacemaker failure related to device manipulation with varying mechanisms, defined by their lead and generator findings on chest X-ray imaging. Reel syndrome is characterized by rotation of the pulse generator on its transverse axis and subsequent lead dislodgement, with the pacemaker lead coiling around the pulse generator.³ We describe a case of a patient with PPM lead dislodgement secondary to Reel syndrome who presented with a misleading clinical picture. We evaluate varying presentations of PPM lead dislodgement, the diagnostic work-up, and practical risk mitigating techniques.

Timeline

First pacemaker procedure (October 2020)	Pacemaker insertion for sinus node dysfunction
Presentation (January 2021)	Admitted to hospital following the radiographic and clinical diagnosis of Reel syndrome
Second pacemaker procedure (January 2021)	<ul style="list-style-type: none"> • Pacemaker deactivation due to dangerous inappropriate pacing • Repositioning of pacemaker leads and secure fixation techniques applied • Unremarkable pacemaker check and discharge from hospital

Case presentation

A 77-year-old overweight female, presented with 4-h history of abdominal pain, abdominal wall, and bilateral lower limb jerking movements after forcefully pulling a blanket towards her with her left arm, whilst supine. She has mild cognitive impairment, a known hiatus hernia and Factor V Leiden deficiency. Her dual-lead PPM was implanted for recurrent syncope, secondary to sinus node dysfunction, 3 months prior to presentation.

On arrival to the emergency department, the patient denied chest pain, features of pre-syncope, or symptoms of cardiac failure. On examination, a contracting or 'pulsating' abdominal wall was observed although the lower limb jerking had ceased on arrival to the emergency department. Laboratory investigations were unremarkable and computed tomography aortogram excluded the suspected initial diagnosis of probably ruptured abdominal aortic aneurysm.

Subsequent chest radiograph (Figure 1A) displayed dislodgement of both pacemaker leads. The right atrial (RA) and right ventricular (RV)

leads had migrated into the superior vena cava and right atrium, respectively (Figure 1A). The patient's electrocardiogram (ECG) revealed inappropriate atrioventricular pacing and failure to capture (Figure 2). The rhythmic, 60 b.p.m., abdominal contractions resolved after PPM deactivation.

The patient was subsequently admitted to the coronary care unit and underwent urgent RA and RV PPM lead repositioning under fluoroscopic guidance. Observations during the procedure suggested the original generator was implanted in the subfascial layer in the left pectoral region and no anchoring stitches were seen in the generator box. The original active pacemaker leads were repositioned and deployed with the optimal numbers of clockwise rotations into the RV apex and RA appendage. This was followed by meticulous suturing of the pacemaker lead sleeves with anchoring stitch to the pectoralis major using non-absorbable sutures. Finally, the pacemaker generator too was anchored to the pectoral muscles using a silk suture. The post-procedure recovery period was complicated by rapid atrial fibrillation, managed with oral metoprolol. Post-procedure chest X-ray showed optimal generator and lead positions (Figure 1B). The patient was discharged the following day. The patient's family was advised about potential re-complications and risk mitigating techniques were provided; active reinforcement of avoiding device manipulation, wearing of a broad arm sling, education about subconscious (i.e., during sleep) device manipulation, and avoidance of excessive shoulder abduction and rotation. Routine cardiology follow-up was arranged.

Discussion

We report on a case presentation of an elderly lady with Reel's syndrome, presenting late after PPM insertion with features suggestive of extra-cardiac wide-spread neuronal stimulation. This presentation occurred 3 months after PPM insertion, which is much longer than expected for lead dislodgement complications, particularly Reel syndrome.⁴ It is likely that the generator had been chronically manipulated in a rotational manner, resulting in the shortening of the PPM leads that was only clinically apparent when the leads began to stimulate extra-cardiac anatomy after vigorous shoulder abduction and arm flexion.

Reel syndrome is a rare cause of device malfunction and is often included in a pool of macro dislocation lead dysfunction secondary to device manipulation, similar to the Twiddler's and Ratchet syndromes. Proposed risk factors for these conditions include female gender, older age, obesity, cognitive impairment, and loose generator implant pocket.⁵ The Reel syndrome is characterized by manipulation and rotation of the generator on its frontal plane, much like the 'reeling up of a coil'. This results in subsequent lead dislodgement, often in the absence of significant lead damage.⁴ Twiddler's syndrome results in lead dislodgement and braiding due to generator rotation on its long axis which may be caused by intentional or unintentional external device manipulation. This condition frequently requires PPM lead replacement because of the effect of torsional force and friction on lead integrity. Ratchet syndrome is caused by 'sliding' of the generator back and forth, laterally along the frontal plane resulting in lead displacement with ratcheting.⁶ One proposed mechanism for lead dislodgement in Ratchet syndrome is the 'unidirectional brake' effect of

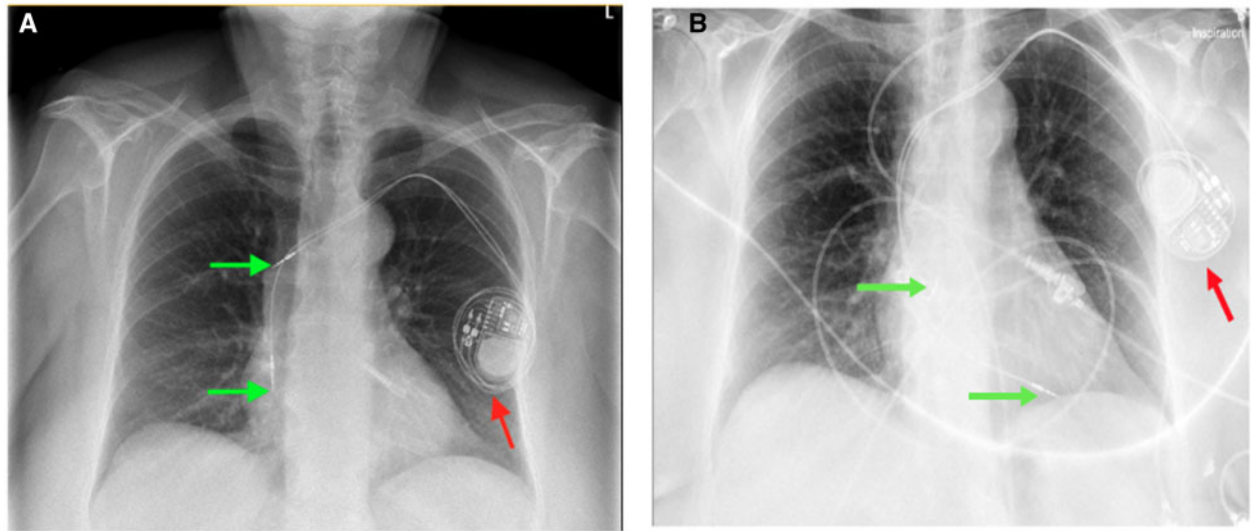


Figure 1 Presenting and post-procedure chest X-ray. Chest X-ray showing dislodged electrodes (green arrow) and leads coiled around the generator (red arrow) (A, left). This chest X-ray reveals characteristic findings of reel syndrome with pacing leads migration and coiling around the pacemaker generator. A repeat chest X-ray (B, right) is performed after permanent pacemaker lead repositioning and reveals adequately placed electrodes (green arrows) and a generator absent of significant coiling (red arrow).

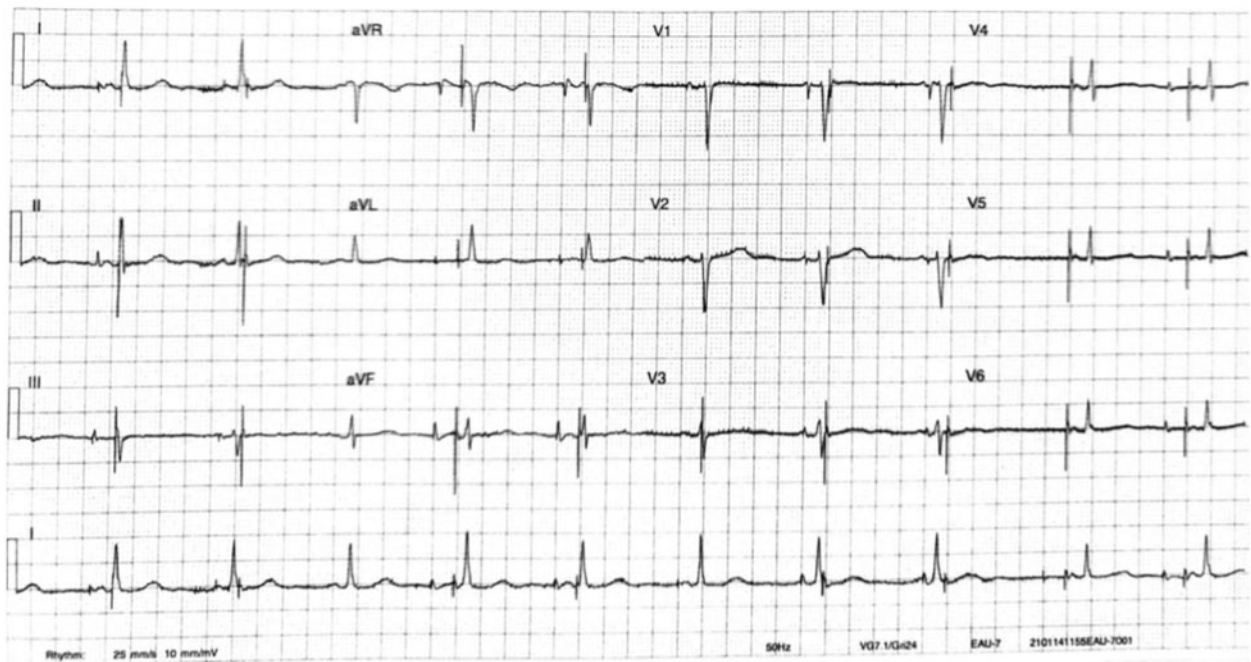


Figure 2 Electrocardiogram at the time of presentation. This electrocardiogram taken in the emergency department reveals intermittent atrial pacing, under sensing of the ventricular lead and ventricular pacing spikes occurring after the QRS complex. The homogenous QRS morphology in the presence of variable ventricular pacing suggests a complete absence of ventricular capture.

a suture sleeve, resulting in a one-way mobilization of PPM leads after device manipulation.⁷ The different radiological manifestation of device manipulations are described in *Figure 3*.

Permanent pacemaker lead dislodgement can present in many unconventional ways. Whilst an exacerbation of cardiac pathology, the reason the device was inserted, is a common cause for presentation,

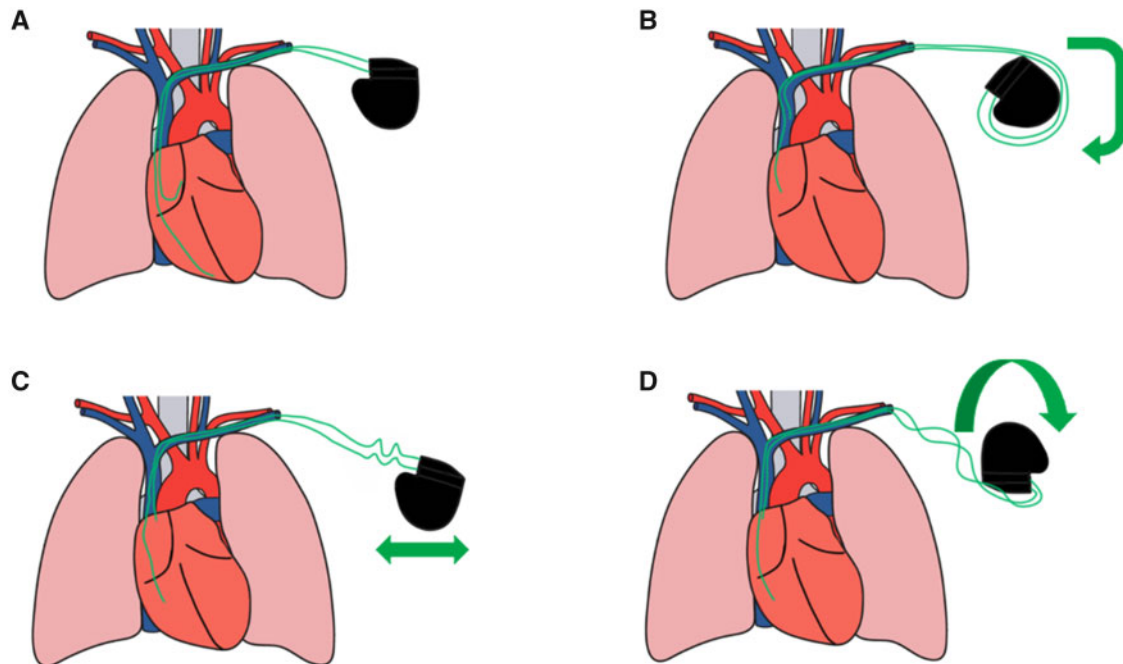


Figure 3 Manifestations of device manipulation-related pacemaker lead dislodgement. This figure depicts the classical radiological and anatomical effects of device manipulated-related permanent pacemaker lead displacement. The normal permanent pacemaker insertion is depicted in panel A. In panel B, the pacemaker generator has been rotated along the frontal plane, resulting in a Reel syndrome and associated lead displacement. Panel C depicts the lead shortening associated with Ratchet syndrome, where the generator is translated back and forth laterally along the frontal plane, resulting in lead displacement. Panel D depicts a classical Twiddler syndrome, where the generator is rotated along the axial plane, resulting in lead dislodgement and braiding. This can often damage the lead integrity and may require permanent pacemaker lead replacement.

patients may present with extra-cardiac manifestations posing a diagnostic dilemma for attending physicians. Rhythmic stimulation of extra-cardiac structures often results in regular contractions of muscles at the same rate as the PPM backup rate. In the case described, the abdominal muscle contractions are strongly indicative of T7 to T11 anterior rami stimulation which causes an activation of the thoracoabdominal nerves, innervating the abdominal wall muscles. Furthermore, the phrenic nerve sits alongside the superior vena cava and stimulation of this nerve may result in rhythmic diaphragmatic contractions, which may manifest as abdominal pulsations as witnessed in the current case. The short-lived bilateral lower limb movements are less easily explained and have yet to be described in the literature, though there are a number of plausible explanations. The PPM lead may have stimulated the sympathetic trunk, resulting in propagation of electric signals to the lumbar plexus, via rami communicantes, resulting in hip flexor activation (i.e. Psoas major). Alternatively, stimulation of the dorsal ramus of the thoracic spinal nerve either directly or via rami communicantes, may result in propagating thoracic spinal contractions, with compensatory hip flexion or extension. Finally, it is possible that high voltage stimulation of thoraco-abdominal nerves resulted in severe rectus abdominus flexion and subsequent posterior rotation of the pelvis, resulting in compensatory leg movements. A pictorial explanation of possible PPM lead displacement presentations is provided in *Figure 4*.

Pacemaker lead dislodgement, whether related or unrelated to device manipulation, can occur in as many as 8% of patients⁸ and may

carry significant mortality, likely related to the underlying arrhythmia requiring PPM insertion; up to 40% at 1 year in those with complete heart block.⁹ For this reason, early identification of lead dislodgement is required to avoid harmful complications to the patient. However, given the somewhat diverse manifestations of PPM lead dislodgement (*Figure 4*), a thorough, stepwise approach should be taken for all patients with cardiac or neuromuscular presentation and a history of PPM insertion. This approach can be divided depending on a patient's presentation; predominantly cardiac or predominantly neuromuscular. A cardiac-dominated presentation, such as chest discomfort, palpitations, pre-syncope or syncope requires a thorough history and examination to exclude acute life-threatening causes such as ST-elevation myocardial infarction, tachyarrhythmia (i.e. ventricular tachycardia), cardiac tamponade or others. Then, a structured approach consisting of ECG, chest X-ray, cardiac monitoring, and pacemaker interrogation is essential. However, a more neuromuscular-dominated presentation should always consider PPM lead dislodgement as a cause and a diagnostic work-up is shown in *Figure 5*. Important differentials for pulsatile movement of the limbs, chest, or abdomen are included in [Supplementary material online, Table S1](#).

Risk mitigating techniques are essential for the prevention of pacemaker lead dislodgement and some techniques are particularly important for the prevention of manipulation-related device complications. Technical tips for reducing PPM lead dislodgement from device manipulation include a sub-pectoral implantation site, a tight fit for the generator pocket and securing PPM leads with non-

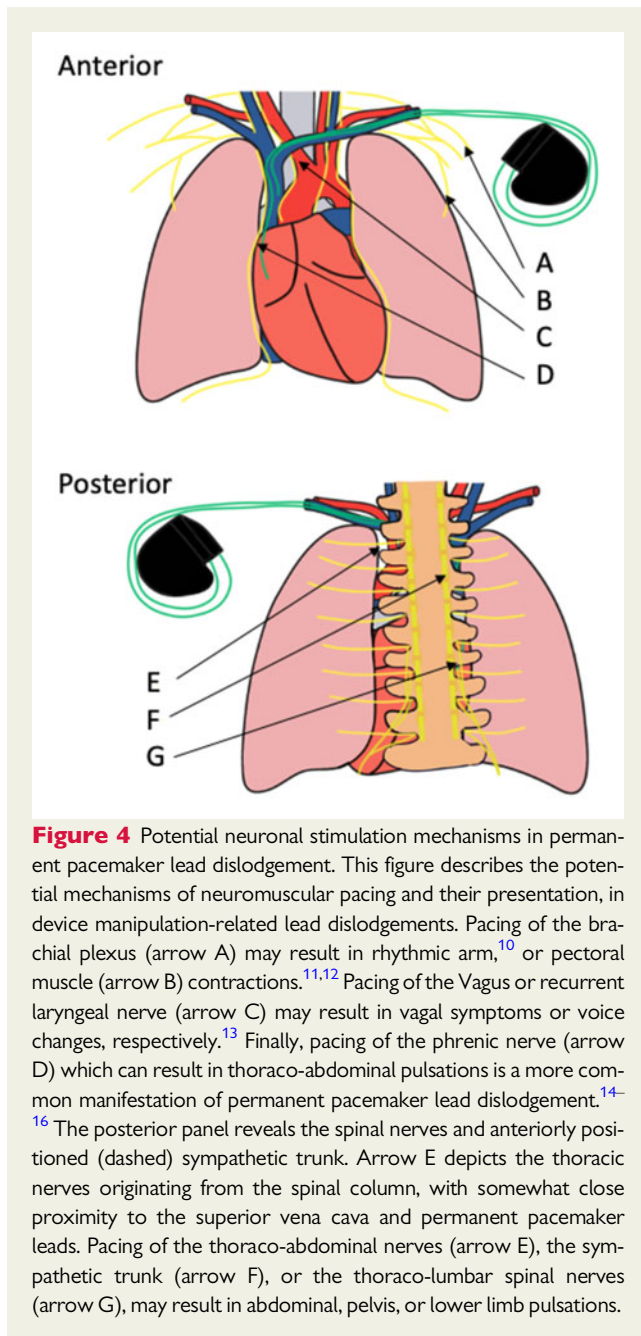


Figure 4 Potential neuronal stimulation mechanisms in permanent pacemaker lead dislodgement. This figure describes the potential mechanisms of neuromuscular pacing and their presentation, in device manipulation-related lead dislodgements. Pacing of the brachial plexus (arrow A) may result in rhythmic arm,¹⁰ or pectoral muscle (arrow B) contractions.^{11,12} Pacing of the Vagus or recurrent laryngeal nerve (arrow C) may result in vagal symptoms or voice changes, respectively.¹³ Finally, pacing of the phrenic nerve (arrow D) which can result in thoraco-abdominal pulsations is a more common manifestation of permanent pacemaker lead dislodgement.^{14–16} The posterior panel reveals the spinal nerves and anteriorly positioned (dashed) sympathetic trunk. Arrow E depicts the thoracic nerves originating from the spinal column, with somewhat close proximity to the superior vena cava and permanent pacemaker leads. Pacing of the thoraco-abdominal nerves (arrow E), the sympathetic trunk (arrow F), or the thoraco-lumbar spinal nerves (arrow G), may result in abdominal, pelvis, or lower limb pulsations.

absorbable sutures via an anchoring stitch technique. The PPM generator can be anchored to the pectoral muscle using non-absorbable sutures and the electrode can be secured via the use of active fixation leads. A firm ‘tug’ at time of PPM lead securing can ensure adequate fixation. Further patient- and family-specific techniques include adequate education about potential complications of PPM insertion and the mechanisms of such, with focus on avoidance of device manipulation. Using a broad arm sling can assist in avoidance of unintentional device manipulation by excessive arm movement. For patients with cognitive impairment, this may be particularly useful during sleep by reinforcing the need to keep the arm adducted and providing a good memory tool to avoid subconscious device manipulation. Furthermore, traditional techniques to avoid a delirium in patients

with cognitive impairment is essential, and this would require adequate perioperative prevention and management of pain, constipation and nosocomial infections. Management of these conditions in the community will also likely reduce unintentional device manipulation in the setting of cognitive impairment and concomitant delirium.

The unique nature of this case reinforces the importance of considering PPM lead dislodgement as the cause for patients presenting with extra-cardiac signs and symptoms, when the aetiology is unclear. The importance of early identification and rapid intervention can be lifesaving, as the loss of pacing function and inappropriate pacing can precipitate dangerous electrophysiological manifestations. All patients with a PPM who present with undifferentiated neuromuscular presentations should be considered to have a PPM lead dislodgement and if this is suspected, undergo an ECG, chest X-ray, and cardiac monitoring until a pacemaker interrogation can be performed.

Lead author biography



George P. Petrov, born in 1994, now working at Joondalup Health Campus, Perth, Western Australia (resident medical officer in cardiology). Graduated from Trinity College Dublin in 2018 and is due to commence surgical training in Ireland, with a speciality interest in cardiothoracic surgery.

Supplementary material

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

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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 images and associated text has been obtained from the patient in line with COPE guidance.

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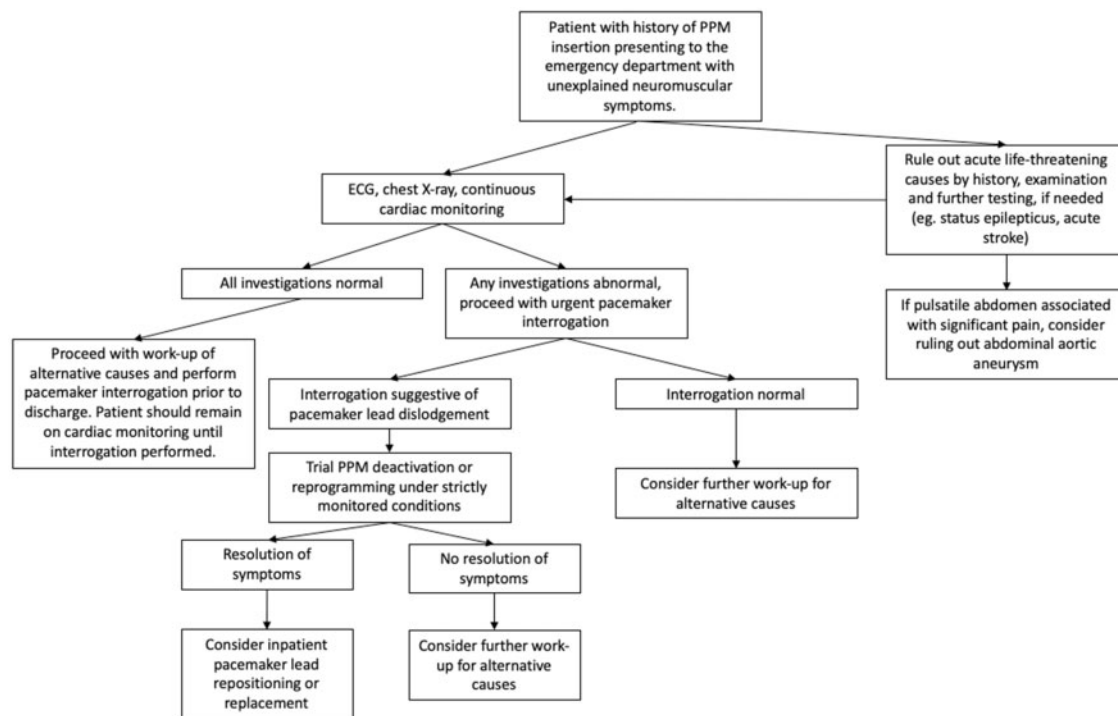


Figure 5 Diagnostic work-up of patients with a history of permanent pacemaker insertion who present with unexplained neuromuscular symptoms. The figure describes a stepwise approach to diagnosing pacemaker lead dislodgement in patients presenting with a history of permanent pacemaker insertion and unexplained neurological symptoms, which may be attributed to permanent pacemaker lead dislodgement. An important first step is to rule out acute neurological causes that may require urgent treatment. All patients should receive an electrocardiogram, chest X-ray, and cardiac monitoring. Then, guided by the presentation and baseline investigations, urgency of pacemaker interrogation can be determined.

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