Twiddler's syndrome and pectoral stimulation in a patient with dual-site unipolar atrial pacing

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Pacing; Sick sinus syndrome.

Twiddler's syndrome is a rare condition of pacemaker rotation; when associated with unipolar pacing it could provoke extracardiac muscular stimulation.

We report a case of an obese woman who unintentionally reversed her triple-chamber, dual-site unipolar atrial pacemaker, implanted for the prevention of paroxysmal atrial fibrillation.

The extracardiac pectoral stimulation was due to unipolar atrial stimulation. The polarity mode was not programmable and output reduction was unsuccessful. Thus, the problem was resolved non-invasively by means of external manual rotation of the pacer. At 24 months of follow-up, the patient was still free of symptoms.

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Introduction

Purposeful or unintentional manipulation of an implanted pulse generator may cause an axial rotation of the pacer and/or twisting of the lead with its eventual fracture or dislodgment. This particular condition has been termed "Twiddler's syndrome"[1].

This relatively rare syndrome was described for the first time in 1968[2]; thereafter, several cases have been reported both in patients implanted with pacemakers and in patients implanted with automatic cardioverter-defibrillators (ICD). Previous reports pertained to abdominal devices[3] but more recently the syndrome has been described for pectoral or even in subpectoral implanted pulse generators[4]. The most important complications of the syndrome include failure of atrial or ventricular sensing, loss of atrial or ventricular capture and extracardiac pacing related to unipolar pacing[5]. In patients with ICD, inappropriate or delayed shocks are relevant issues; in 1994, a case of sudden cardiac death was reported in a patient who had been implanted with an ICD[6].

We here report the case of a woman with a pectoral implanted multisite unipolar atrial DDDR pacemaker who presented with right pectoral and deltoid muscle stimulation due to axial rotation of the device.

Case report

A 61-year-old woman with brady-tachy syndrome was admitted to our institution because of recurrent and symptomatic (palpitations and dyspnea) episodes of paroxysmal atrial fibrillation, resistant to propafenone, amiodarone and flecainide therapy. Baseline ECG showed an increased P-wave duration (Fig. 1); at two-dimensional echocardiography, the left atrial dimensions were normal; at 24-hour ECG monitoring, several episodes of paroxysmal atrial fibrillation with significant pauses after spontaneous restoration of sinus rhythm were observed.

The patient was obese (body mass index 31.25 kg/m^2) and used to help herself with crutches, because of an invalidating spondylo-arthrosis.

We implanted a multisite atrial DDDR pacemaker (Chorum 7337 MSP, Ela Medical, Le Plessis-Robinson, France) connected to: 1) a bipolar lead (Stelid BT46D; Ela Medical, France) in the apex of the right ventricle; 2) a bipolar lead (Stelid BJ45D; Ela Medical, France) in the right auricle; 3) a screw-in unipolar lead (Tendril 1388K; St. Jude Medical, Sylmar, CA, USA) at the ostium of the coronary sinus (Fig. 2). The pulse generator was placed subcutaneously in the right pectoral region (Fig. 3A); the can, the right ventricular and right auricular leads (inserted through the right cephalic vein) were not anchored to the muscle fas-

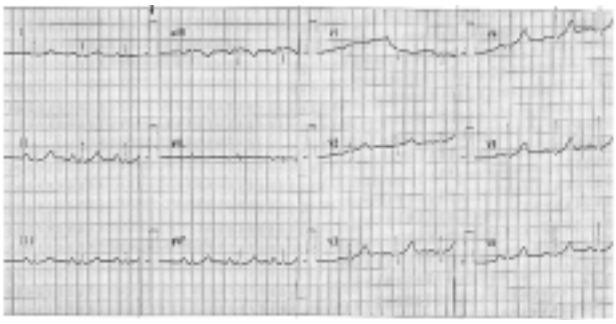

Figure 1. *Twelve-lead baseline ECG shows an increased P-wave duration.*

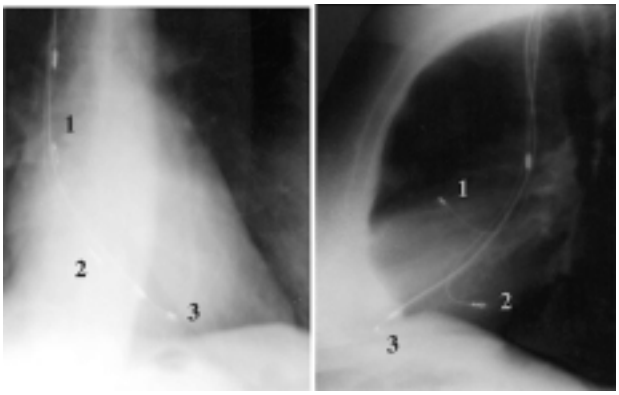

Figure 2. *X-ray lead position for multisite atrial DDD pacing* (left: *antero-posterior view;* right: *latero-lateral view). 1 = bipolar J lead in the right auricle; 2 = screw-in unipolar lead at the ostium of the coronary sinus; 3 = bipolar right ventricular lead.*

cia; only the unipolar lead placed at the ostium of the coronary sinus (inserted through the right subclavian vein) was fixed to the underlying subcutaneous fascia. The Chorum 7337 MSP is a triple-chamber pacemaker with one right bipolar atrial site, one left unipolar atrial site and one unipolar ventricular site. Atrial pacing and ventricular sensing/pacing are unipolar and not programmable. The pacemaker's titanium case acts as the anode and the distal electrodes (ventricular and two atrial ones) act as the cathode; atrial sensing is programmable in bipolar configuration[7].

The patient was discharged with the pacemaker programmed in DDTA mode (multisite unipolar atrial stimulation) and on flecainide (200 mg/die) and nadolol (40 mg/die) therapy.

After 2 months of satisfactory pacemaker function, the patient returned to our institution complaining of recurrent right pectoral and deltoid muscle stimulation.

On admission, the body mass index was unchanged, the wound was well-healed and the pocket size seemed not to be much greater than the size of the pulse generator. The ECG showed correct multisite atrial stimulation without signs of atrial or ventricular sensing or pacing failure (Fig. 4).

Chest X-ray showed that the leads were well positioned and without evidence of dislodgment or twisting; nevertheless, the pulse generator identification code was oriented inferiorly while at immediate postoperative study it was correctly oriented superiorly, suggesting an axial rotation of the can with consequent exposure of the uninsulated side of the device to the right pectoral muscle (Fig. 3B).

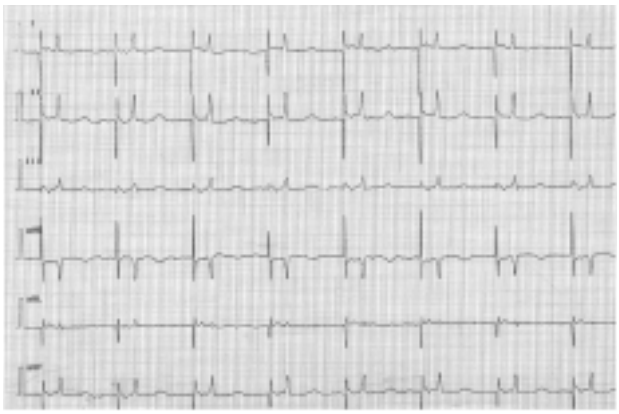

Figure 4. *Correct atrial multisite unipolar pacing with a narrow P wave (peripheral ECG derivations).*

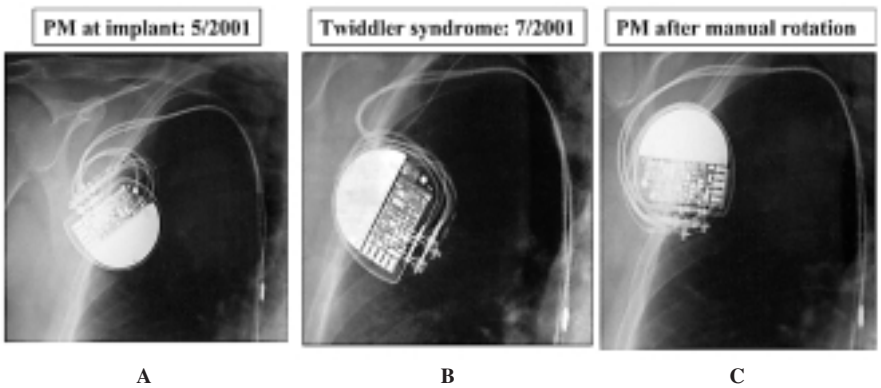

Figure 3. *X-ray position of the pacemaker (antero-posterior view).* A: *original position of the pacemaker after implant (May 2001) with correct orientation of the lead connectors.* B: *Twiddler's syndrome after 2 months (July 2001) with rotation of the pacemaker case.* C: *pacemaker after restoration of the original position by external manual rotation of the case.*

To exclude that the ventricular lead was responsible for the clinical symptoms, the pacemaker was temporarily programmed in VVI mode at high output voltage (5 V, 0.49 ms): no extracardiac stimulation occurred.

With the pacemaker reprogrammed in AAI (right atrial and low interatrial septum unipolar pacing) and high output voltage (5 V, 0.49 ms) relevant pectoral and deltoid contractions were observed; a slight decrease in the energy output (4 V, 0.49 ms) resulted in the loss of deltoid contraction with persistent right pectoral stimulation; with a further decrease of the output voltage (3 V, 0.49 ms), no more extracardiac stimulation was noted. As the multisite atrial stimulation threshold was satisfactory (1.5 V, 0.49 ms), the pacemaker was programmed in DDTA with an atrial energy output of 2.5 V (0.37 ms).

After few hours, the patient again complained of mild right pectoral contraction. Thus, we decided to try to revolve the pulse generator externally without any surgical revision: with the patient mildly sedated and under fluoroscopic monitoring, the can of the pacemaker was successfully rotated manually on its axis (Fig. 3C); its position seemed stable even during extreme right arm movements and no compressive dressing was necessary.

The patient was followed closely and at 24 months she is still free of extracardiac stimulation and the system functions well. Pacemaker telemetry showed only brief periodic episodes of paroxysmal atrial fibrillation, concordant with sporadic palpitations of the patient.

Discussion

Twiddler's syndrome is a rare but relevant clinical syndrome with various clinical manifestations; the patient may be asymptomatic or, more often, complains of symptoms related to pacemaker or defibrillator malfunction with potentially fatal complications[1-6].

Elderly patients and obese women, with abundant subcutaneous fat or atrophy of the subcutaneous tissues which allow for rotation and torsion of the pulse generator in the pocket, seem to be at higher risk[8,9]. However, the syndrome has also been observed in young individuals[10,11].

Among the possible complications of the syndrome, local muscle stimulation is rare. With older unipolar devices muscular contractions in the late post-implant period were probably due to erosion of the pacemaker's protective coating and/or lead fracture; but with current bipolar pacemakers, pectoral muscle stimulation is very unusual.

At present, unipolar stimulation is used in multisite pacing, for atrial or ventricular resynchronization. To our knowledge, this is the first case of Twiddler's syndrome in multisite (atrial) pacing to be reported.

Various preventive methods, such as fixation of the electrodes and/or pacemaker can to the surrounding tissues[12], preparation of an appropriate size pocket, implant of the pulse generator subpectorally or its insertion in a Dacron pouch, have been proposed[13]. Nevertheless, these measures cannot completely eliminate Twiddler's syndrome: only bipolar stimulation could prevent extracardiac muscular stimulation in this clinical situation[14].

Just as the majority of the patients in which this syndrome has been described, the patient we report upon was a middle-aged obese woman; the obesity was also complicated by an important spondylo-arthrosis with the use of crutches and recurrent unintentional manipulations on the pacemaker pocket. In the present case, we observed only the rotation of the pacemaker can without any dislodgment of the pacing leads; the anchorage of the pulse generator to the underlying fascia with non-absorbable sutures could be useful in preventing this complication in patients considered at risk for this syndrome. However, in this case the problem could be corrected non-invasively, with psychological assistance and manual painless external rotation of the pacemaker case.

In conclusion, pacemaker or ICD Twiddler's syndrome is a rare but clinically relevant complication which must be suspected in the presence of malfunctioning systems; programmed complete evaluations of adequate functioning of the device, including pocket evaluation and chest radiography in standard projections, are mandatory because this potentially lethal complication may be easily detected by means of radiographic examination.

References

1. Furman S, Hayes DL, Holmes DR. A practice of cardiac pacing. 2nd edition. Mount Kisko, NY: Futura Publishing Company, 1989: 499.
2. Bayliss CE, Beanlands DS, Baird RJ. The pacemaker-Twiddler's syndrome: a new complication of implantable transvenous pacemakers. Can Med Assoc J 1968; 99: 371-3.
3. Guharay BN, Ghose JC, Majumdar H, Basu AK. The pacemaker-Twiddler's syndrome: another disadvantage of abdominal implantation of pulse generators. Br J Surg 1977; 64: 655-60.
4. Kumar A, McKay CR, Rahimtoola SH. Pacemaker Twiddler's syndrome: an important cause of diaphragmatic pacing. Am J Cardiol 1985; 56: 797-9.
5. Ciolli A, Di Lorenzo M, Lo Sardo G, Palamara A. Pacemaker Twiddler's syndrome: an unusual form of pacing wire displacement. Ital Heart J 2001; 2: 321-2.
6. Avitall B, Stormo A, Barragry T, Axtel K, Hare J. Sudden cardiac death: Twiddler's syndrome with an implantable cardioverter-defibrillator. Am Heart J 1994; 128: 833-6.
7. Chorum 7337 MSP. Physician's manual. Le Plessis-Robinson: Ela Medical, 1998.
8. Khalilullah M, Khanna SK, Gupta U, Padmavati S. Pacemaker Twiddler's syndrome. A note on its mechanism. J Cardiovasc Surg 1979; 20: 95-100.
9. Fahraeus T, Hoijer CJ. Early pacemaker Twiddler syndrome. Europace 2003; 5: 279-81.
10. Berul CI, Hill SL, Estes NA. A teenager with pacemaker Twiddler syndrome. J Pediatr 1997; 131: 496-7.

11. Servetar EM. A case report of Twiddler's syndrome in a pediatric patient. J Pediatr Oncol Nurs 1992; 9: 25-8.
12. Stanford W, Coyle FL, Dooley BN, et al. Transvenous pacemaker failure: migration of catheter lead by patient manipulation. Ann Thorac Surg 1968; 5: 162-4.
13. Parsonnet V, Bernstein AD, Neglia D, et al. The usefulness of a stretch-polyester pouch to encase implanted pacemakers and defibrillators. Pacing Clin Electrophysiol 1994; 17: 2274-8.
14. Said SA, Van Driel BA, Onrust A. Pacemaker Twiddler's syndrome: delayed transection of permanent ventricular lead. Eur Heart J 1996; 17: 1914-5.