

Twiddler Syndrome: A Rare Complication with Important Clinical Relevance

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ABSTRACT

Twiddler syndrome is a rare complication observed after implantation of pacemakers and ICDs due to conscious or unconscious "twiddling" at the implantation side with the result of device malfunction. In patients at risk for all solutions proposed in literature to date (active fixation, small pockets, subpectoral implantation, etc.) have been reported cases of failure. We describe a new technique based on additional suture of the leads. In our experience it revealed to be very useful and we think that it could be applied with success in patient at risk for life.

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Key words: Twiddler syndrome; Pacemaker malfunction; Leads dislodgment

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INTRODUCTION

Twiddler Syndrome is a rare cause of implanted device (pacemaker or ICD) malfunction^[1-3]. It is a condition due to the conscious or unconscious manipulation of the case at the implant side which cause

its rotation into the pocket^[3,4]. Generator's rotation around himself causes winding and dislodgment of the leads with a mechanism in which the final causes are not yet totally known^[5]. However these are crucial for the device malfunction^[6]. The few experiences in literature show that this condition is common in psychiatric^[7-8] and pediatric^[9,10] patients as well as elderly with greater laxity of the tissues and in the presence of obese subjects^[4,10-12]. More generally this condition is found in patients with cognitive impairment. Typical symptoms can be related to the loss of capture and of sensing with consequent hypoperfusion (tiredness, fatigue, confusion, presyncope and syncope)^[8]. Less frequent, but always possible, symptoms are related to the damage of the nearby organs^[4,13]. Have been reported sensation of diaphragmatic and abdominal pulsation due to the ipsilateral phrenic nerve stimulation or rhythmic arm movements due to brachial plexus stimulation^[12]. However there are also cases without clinical symptomatology^[14]. In ICD implanted patients it can be the cause of inappropriate shocks and malfunction^[15-16].

DISCUSSION

Twiddler syndrome was first identified by Bayliss in 1968^[11]. From that period in literature have been published several but sporadic case reports. The first experiences were based on loss of sensing and of capture^[1,17,18]. The subjects that showed this condition were united by cognitive impairment, especially psychiatric and elderly as well as pediatric patients. This obvious kind of manifestation however it's not the only. In fact there are reported also cases of isolated atrial lead fault sensing in presence of normal ventricular capture^[16-19] in dual chamber pacemaker. Even has been reported a unique case in which Twiddler syndrome caused hiccups and dysphonic metallic voice^[20]. In ICD and CRT-D implanted patients the main manifestations are inappropriate shocks^[15]. In major risks are involved patients with syncope or aborted sudden cardiac death due to not cardioverted sustained ventricular tachycardia. All this manifestation can occur in months with the device twiddling also if has been reported a case in which the first exit block appeared after only 17 hours^[21]. The mechanic displacement of the device could find other "facilitating" causes. One of the facilitating factors may be the weakness of the tissue associated with an oversized pocket. Incidence of the problem probably has grown up in the last years with the use of smaller devices^[22]. In almost all cases of manifestation of Twiddler Syndrome

there is a history of manipulation, by the patient, in correspondence of the pocket, with feedback at the same site of injury from scratching. It is assumed that the action of scratching, acting on predisposing factors (tissue laxity, large pockets etc.), determines the rotation of the device body with stretching and dislodgement of the leads which probably rewind on their own. Another hypothesis is based on the fact that scratching involves a stretching of leads which are so displaced and rewound on themselves.

To date the main solutions are represented, in patients at risk, by the use of leads with helical screws (active fixation)^[23], subpectoral implantation and formation of small pockets^[4,24]. There however are cases of failure of these solutions^[25-27].

CONCLUSIONS

Twiddler syndrome is a potential risk in special populations of patients who need a device implantation. Particular risk is observed in pacemaker dependent subjects as well as those with an ICD implanted (inappropriate shock and sudden cardiac death risk)^[28]. For this reason, although it is a rare condition, it is important to keep in mind its high clinical importance. To date for all the main solutions proposed for patients at risk (active fixation, small pockets, subpectoral implantation, etc.) are reported cases of failure.

Our proposal, in the case of patients at risk (and between them especially in those who are pacemaker dependent), is to practice implantation exactly at the level of the pectoral plan, fixing the case of the pacemaker to the muscle, and to make a further leads' fixation (beyond the normal suture of the lead close to the entry into the cephalic and/or subclavian vein) to the pectoral. We propose that other leads' fixations should be made with repair sleeves (used for repairing in leads' micro-loss of insulation). In fact these devices are provided with eyelets, usable for further leads' anchoring to the muscle.

With this kind of setting, it will be very difficult for conscious or unconscious manipulation to determine rotations of the pacemaker body on itself into the pocket and/or the stretching of the catheters.

In our experience, even if in a single case with DDD pacemaker, this technique provided a solid anchor and, at the 6th month follow up, even in the presence of objective manipulation attempt (marked lesions from scratching at the implantation site), telemetry control and X-Ray showed no dislocation of the lead.

Even if it is not a known technique besides being time consuming it could be useful also for patients at risk of life.

CONFLICT OF INTERESTS

There are no conflicts of interest with regard to the present study.

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