Twiddler’s Syndrome Complicating a Transvenous Defibrillator Lead System*

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Twiddler’s syndrome is a rare complication seen in patients with implanted pacemakers or defibrillators. The condition typically presents with device malfunction and occurs when the patient either consciously or unconsciously twists and rotates the implanted device in its pocket, resulting in torsion and dislodgement of the implanted lead. A case of twiddler’s syndrome involving a transvenous defibrillation lead and an abdominally implanted defibrillator is described. This is the first report of this complication with this particular lead. The patient in this report was a middle-aged obese diabetic woman who presented 7 months after defibrillator implantation with device noncapture and intermittent nonsensing. Review of the literature reveals that the majority of patients with this complication are middle-aged obese women with a defibrillator pocket that exceeds the size of the defibrillator. Treatment measures are discussed both for the patient with this complication and for the patient at increased risk for its occurrence. (CHEST 1996; 109:1391-94)

Key words: implantable defibrillator; twiddler’s syndrome; ventricular tachycardia

Abbreviations: ICD=implantable cardioverter defibrillator; VT=ventricular tachycardia

Pacemaker twiddler’s syndrome is a rare complication of permanent pacemaker implantation which typically presents with pacemaker malfunction. The condition occurs when the patient, either consciously or unconsciously, rotates or “twiddles” the implanted pacemaker in its pocket, resulting in torsion, dislodgement, and often fracture of the pacing lead. The diagnosis is confirmed by a chest radiograph which reveals a twisted, entangled, and dislodged pacing lead. The condition, which was first described by Bayliss et al in 1965, can occur if the subcutaneous tissues are lax, if the pacemaker is untethered in its pocket, or if the size of the pacemaker pocket exceeds that of the pacemaker. Defibrillator twiddler’s syndrome is a more modern variant of this condition in which the implanted device is an implantable cardioverter defibrillator (ICD) rather than a pacemaker. Although there are a number of reports of defibrillator twiddler’s syndrome with epicardial defibrillator systems, the present report is, to our knowledge, the first to describe its occurrence with an exclusively transvenous ICD system.

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A 54-year-old woman presented with recurrent episodes of congestive heart failure. During her hospital admission, she was noted to have long runs of unimorphic ventricular tachycardia (VT) at a rate of 175/min, which was well tolerated. She denied any medical history of syncope or presyncope. Her VT was suppressed with intravenous lidocaine and her heart failure was treated successfully with digoxin, furosemide, and captopril. Although she had no definite history of myocardial infarction, she had significant risk factors for coronary artery disease including long-standing insulin-dependent diabetes mellitus, hypercholesterolemia, cigarette smoking, and morbid obesity. Examination revealed a markedly obese woman with a body weight of 250 lb and a height of 168 cm.

The patient underwent cardiac catheterization which demonstrated significant left ventricular dysfunction with a left ventricular ejection fraction of 0.35. The right coronary artery was occluded. There was diffuse noncritical disease in the left anterior descending coronary artery and the distal circumflex/obtuse marginal artery displayed a recent occlusion which was successfully reopened with angioplasty. Her baseline electrophysiologic study demonstrated inducible sustained unimorphic VT of two morphologies (right bundle branch block, inferior axis, cycle length—240 ms; and left bundle branch block, superior axis, cycle length—240 ms) induced with three extrastimuli and terminated with direct current shock. There was also sinus node dysfunction which was aggravated by intravenous procaainamide hydrochloride. After discussion of the various options with the patient and her family, it was decided to implant a nonthoracotomy or transvenous ICD with the capacity to provide backup bradycardia pacing and noninvasive programmed ventricular stimulation.

An endocardial defibrillation lead (Cardiac Pacemakers, model 0064, Endotak C) was advanced to the right ventricle via the left subclavian vein, and the lead tip was positioned at the low interventricular septum. The final pacing parameters consisted of a threshold of 0.5 V, a current of 0.5 mA, a resistance of 400 ohms, and an R wave of 19 mV. Defibrillation threshold testing revealed a lead-alone monophasic defibrillation threshold of 20 J or less. The lead was secured in position in the left infraclavicular fossa with a strain-relief loop in place, and the lead was tunneled to a suprafascial defibrillator pocket in the left hypochondrium. The lead was connected to a Medtronic Pacer-Cardioverter-Defibrillator (model 7217B), and induced ventricular flutter was sensed appropriately and terminated with a single 34-J shock. The defibrillator and lead were buried in the pocket which was closed in three layers. The patient had an uneventful postoperative recovery, and posteroanterior and lateral chest radiographs (Fig 1) confirmed that the lead was in a satisfactory position. Follow-up defibrillator evaluation in the Electrophysiology Laboratory prior to discharge home confirmed normal function with a VT cardioversion threshold of 18 J or less. The defibrillator was programmed for VT detection at a cycle length of 350 ms with the 1st therapy being an 18-J shock and the 2nd through 4th therapies being 34-J shocks. Ventricular fibrillation detection was programmed at a cycle length of 320 ms, and all 4 VF therapies were 34-J shocks. Ventricular demand pacing was programmed at 40 beats per minute, with output at 5.4 V, pulse width of 1.59 ms, and sensitivity of 0.3 V. The patient was discharged home well and returned to the ICD clinic at 3 and 6 months afterward for routine follow-up evaluations. She reported no symptoms. Interrogation of the device revealed no VT or ventricular fibrillation episodes or therapies. The pacing threshold was at a pulse width of 0.06 ms at 5.4 V and at a pulse width of 0.15 ms at 2.8 V. Sensing was appropriate at 0.3 mV apart from occasional postspacing T wave oversensing which had been found on previous evaluations.

The patient was admitted to the hospital 7 months after ICD implantation for treatment of decompensated congestive heart failure. On routine telemetry monitoring, the ICD displayed intermittent nonsensing and noncapture. Programmer analysis proved difficult due to the patient’s obese abdomen but confirmed ventricular noncapture even at the maximum output and intermittent nonsensing. Posteroanterior and lateral chest radiographs (Fig 2) revealed a significant change in the lead position compared with that in the immediate postoperative study. The Endotak lead was now partially retracted from the right ventricle, the strain-relief loop was no longer present, and the extrathoracic portion of the lead was coiled and twisted repeatedly on itself typical of that seen in twist-diller’s syndrome. On direct questioning, the patient stated that she had never intentionally manipulated the ICD or the lead. However, she stated that occasionally on waking in the morning she noticed the ICD sitting on its side in the abdominal pocket. The problems...
with the lead and ICD could not be corrected noninvasively, and therefore the patient was scheduled for surgical exploration and lead revision.

With the patient under general anesthesia, an incision was made over the ICD pocket in the left hypochondrium. The capsule was incised, and it was clear that the dimensions of the pocket exceeded those of the defibrillator. The ICD was mobilized, and the Endotak lead was found to be tightly coiled and twisted repeatedly on itself in the ICD pocket (Fig 3). Attempts to disentangle the lead were unsuccessful, and the lead was mobilized surgically in the left infraclavicular fossa. Close inspection of the lead revealed that the insulation was interrupted at one point and therefore this lead was abandoned. The lead was then removed intact from the heart. Implantation of a new endocardial defibrillation lead was then performed.

**Discussion**

Defibrillator twiddler's syndrome was first described by Veltri et al in 1984 in a patient with a superior vena cava spring electrode, a left ventricular epicardial patch, and an abdominally implanted ICD. Since then, there have been at least four other case reports of defibrillator twiddler's syndrome in patients with epicardial lead systems and abdominally implanted defibrillators. Four of the five patients in these reports were women aged 38, 51, 53, and 58 years old, and the fifth patient was a 45-year-old diabetic man. Four of the patients were described as being obese, and in two, the defibrillator pocket was described as being larger than the defibrillator. In 2 patients, the problem was discovered at the time of elective replacement of an end-of-life unit; 1 patient presented 4 months after elective replacement of an end-of-life unit and 2 patients presented with device nonfunction 7 to 8 months after defibrillator implantation.

Our patient had many features similar to those of the previous examples of defibrillator twiddler's syndrome. She was a 54-year-old obese diabetic woman who had undergone de novo defibrillator implantation 7 months earlier. She presented with ventricular noncapture and intermittent nonsensing, and a chest radiograph demonstrated the typical withdrawal and entanglement of the endocardial lead. Surgical exploration revealed that the size of the pocket exceeded the size of the defibrillator, thus permitting torsion of the lead and defibrillator.

Various therapeutic approaches have been proposed. The simplest solution involves securing the defibrillator to the rectus fascia with large nonabsorbable sutures and obliterating the redundant fibrous capsule to prevent defibrillator migration and torsion. Other investigators have proposed implanting the defibrillator in a pouch made of a synthetic polyester fiber (Dacron) which is then secured to the fibrous capsule. In some cases, each lead is individually secured to the tissues. Others have advocated implanting and anchoring the defibrillator underneath the rectus muscle anterior to the posterior rectus sheath. The lead or leads should be examined carefully and, as in our case, may need to be replaced.

In conclusion, defibrillator twiddler's syndrome is an infrequent, serious, and potentially fatal late complication of
defibrillator implantation. Prior reports have described its occurrence with epicardially implanted hardware, but our report is the first in a patient with an exclusively transvenous defibrillation lead. Our patient was typical of other patients with this complication in that she was a middle-aged obese woman in whom the size of the defibrillator pocket exceeded the size of the defibrillator. Patients with this profile undergoing defibrillator implantation should be considered at risk for this complication and steps should be taken at the time of implantation to prevent its occurrence. These steps might include anchoring the defibrillator and possibly the lead to the underlying fascia with nonabsorbable sutures, inserting the defibrillator in a Dacron pouch, and taking care not to oversize the defibrillator pocket.

REFERENCES
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Key words: hypertension; hypertrophic cardiomyopathy; pheochromocytoma

Abbreviations: HOCM=hypertrophic obstructive cardiomyopathy

The cardiovascular manifestations of pheochromocytoma are many and varied. Specific cardiac complications include left ventricular hypertrophy, catecholamine-induced myocarditis, and dilated cardiomyopathy, both the latter being potentially lethal conditions. Rarely, the left ventricular hypertrophy associated with pheochromocytoma simulates that of hypertrophic obstructive cardiomyopathy (HOCM). We report two cases in which this complication occurred and show the degree of resolution of these abnormalities after tumor removal.

CASE REPORTS

Case 1
A 44-year-old black woman was admitted to Baragwanath Hospital in 1991 with a diagnosis of suspected pheochromocytoma. She had a 10-year history of hypertension for which she received intermittent treatment. In 1990, she was diagnosed as having HOCM and was treated with atenolol and verapamil. A year later, she developed severe dyspnea and weakness and was found to be ill looking, mildly pyrexial, with a BP of 220/140 mm Hg. There was clinical evidence of marked left ventricular hypertrophy associated with a grade 3/6 ejection systolic murmur heard along the left parasternal border that increased in intensity in the erect position.

Retinal examination revealed grade 2 hypertensive changes. Wide fluctuations in BP were noted on a 24-h ambulatory BP recording with maximum systolic and diastolic BPs of 222 and 132 mm Hg, respectively, and minimum systolic and diastolic BPs of 107 and 75 mm Hg, respectively. On further questioning, the patient revealed that she had experienced repeated episodes of headache, sweating, heat intolerance, palpitations, and vomiting since 1987. The chest radiograph showed left ventricular enlargement; the ECG showed voltage criteria for left ventricular hypertrophy with marked ST-segment and T-wave changes in the inferior and precordial leads. The echocardiographic features are shown in Table 1. Blood investigations showed a neutrophil leucocytosis of 34.3×10⁹/L, and a serum potassium level of 2.9 mEq/L. The plasma norepinephrine level was significantly elevated at 4.175 pg/mL. Abdominal CT scan

Pheochromocytoma Associated With Clinical and Echocardiographic Features Simulating Hypertrophic Obstructive Cardiomyopathy*

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Left ventricular hypertrophy simulating hypertrophic obstructive cardiomyopathy is a rare complication of pheochromocytoma. In this report, two cases of pheochromocytoma with this complication are described. Successful tumor removal in both cases led to relief of symptoms, normalization of BP, regression of abnormal clinical features, normalization of the ECGs, but only partial regression of the echocardiographic features despite prolonged follow-up of 24 and 32 months, respectively. (CHEST 1996; 109:1394-97)

Table 1—Comparison of Two-Dimensional Echocardiographic and Doppler Flow Study Features Before (B) and 32 Months After (A) Removal of Pheochromocytoma in Patient 1*

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<tr>
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<th>B</th>
<th>A</th>
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<tbody>
<tr>
<td>LV posterior wall thickness in diastole, mm</td>
<td>12.3</td>
<td>12.4</td>
</tr>
<tr>
<td>IVM thickness in diastole, mm</td>
<td>18.2</td>
<td>12.4</td>
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<tr>
<td>SAM</td>
<td>Grade 3</td>
<td>Absent</td>
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<tr>
<td>Mitral regurgitation</td>
<td>Mild</td>
<td>Absent</td>
</tr>
<tr>
<td>Mean LV outflow gradient, mm Hg</td>
<td>45</td>
<td>7.9</td>
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<tr>
<td>EF, %</td>
<td>85</td>
<td>79.2</td>
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*EF=ejection fraction; IVM=interventricular septum; LV=left ventricle; SAM=systolic anterior motion of the mitral valve.

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