

Country cardiograms case 63: Answer

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The electrocardiogram (ECG) shown in Fig. 1 (on page 86) reveals third-degree atrioventricular (AV) block with an underlying nonconducting sinus rhythm at a rate of about 80 beats/min. The narrow complex escape rhythm most likely represents a junctional escape rhythm at a rate of 30 beats/min. There was underlying nonconducted atrial activity. Pacing spikes are also clearly visible, at a rate of 75 beats/min, with no ventricular capture (i.e., pacemaker noncapture). The chest radiograph shown in Fig. 2, A (on page 87) shows the pacemaker immediately after implantation, about 2 weeks before the current presentation. A single pacemaker lead is visible in the right ventricle, with the pacemaker pulse generator in the left infraclavicular area. Fig. 2, B is the radiograph at the time of the current presentation. It shows the pacemaker lead retracted, with its tip no longer in the right ventricle. The lead tip is most likely in the right atrium. The device also appears to have migrated laterally and caudally, likely secondary to device manipulation. The patient was admitted to the coronary care unit for monitoring and management.

High-degree AV block is a second-degree AV block in which the P:QRS ratio is 3:1 or higher. Unlike third-degree heart block, there is still some relation between the P waves and the QRS complexes. This produces a very slow ventricular rate. Complete heart block occurs when there is complete electrical dissociation between the atria and the ventricles, with the ventricles beating independently of the atria. This shows as complete AV dissociation on an ECG.

Patients with high-degree AV block and third-degree AV block accompanied by a slow escape rhythm often present with symptoms that include shortness of breath, near syncope and syncope. Symptoms of heart failure and angina may be worsened by the slow ventricular rate. The causes of high-degree and third-degree AV block are diverse.¹ Both high-degree AV block and complete heart block warrant insertion of a transvenous pacing system to prevent symptoms from bradycardia.

Pacemaker lead dysfunction is relatively common within the first year after device implantation, with lead re-intervention required in over 4% of patients.² In 1 large prospective registry, the most common problems encountered were lead dislodgement, malfunction and/or perforation.² The lead most commonly involved was the right atrial lead, and the RV lead was least commonly involved.

Twiddler's syndrome is an uncommon complication after implantation of a permanent pacemaker. It was first noted by Bayliss and colleagues³ in 1968. This syndrome refers to pacemaker malfunction owing to external manipulation of the pacemaker pulse generator by the patient. The patient consciously or subconsciously moves the pulse generator in the pacemaker pocket. In the classic form of the syndrome, this results in the device's spinning on itself, with resultant coiling of the lead(s) in the pocket. When the slack on the lead is removed, further manipulation results in dislodgement of the pacemaker lead from the cardiac chamber. If the ventricular lead dislodges, this often results in cessation in ventricular pacing. This can have serious, life-threatening consequences if

the patient is pacemaker dependent. With continued spinning or reeling of the device in the pocket, further retraction of the lead occurs. If retracted far enough, diaphragmatic pacing may occur, resulting in abdominal pulsations from ipsilateral phrenic nerve pacing. Last, ipsilateral arm twitching can occur from stimulation of the brachial plexus in the arm, in the area of the pacemaker pocket.⁴

Twiddler's syndrome is more likely to occur in young children and older adults. It has also been noted to have an association with obese patients, those with intellectual disabilities and those with dementia.^{5,6} The syndrome is also known to occur in patients with transvenous implantable cardioverter defibrillators, which can result in inappropriate shocks and even defibrillator failure.^{7,8}

Although pacemakers are often implanted at larger hospitals, many patients are from smaller, often remote, communities. As the patient's time in hospital is often short, device malfunction may occur after the patient has been discharged and returned to his or her community. This makes recognizing and diagnosing pacemaker malfunction important for all acute care physicians. Diagnosis of pacemaker malfunction starts with a good symptom history and an understanding of the original device indication. Twelve-lead ECG can be very helpful in identifying rhythm disturbances, particularly when the ECG is compared to a previous ECG. Also essential in the diagnostic investigation is chest radiography. It is essential to obtain both anteroposterior and lateral views in order to fully evaluate the lead position. For the nonexpert, it is best to compare the current radiograph to the radiograph that was done immediately after implantation. One should compare the position of the lead, particularly the distal lead tip, looking for migration. One should also examine the lead along its entire length to assess for fracture, which would appear as a break in the lead's continuity. Finally, the lead-generator connection should be assessed. Again, it is important to compare new and old radiographs and evaluate the contact between the proximal end of the lead and the generator connector terminal, looking mainly for differences.

Acute management of pacemaker malfunction, including twiddler's syndrome, depends on the presentation of the patient. In patients who are pacemaker dependent, dislodgement of a ventricular

pacing lead can result in pacing failure, with extreme bradycardia or asystole. These cases warrant urgent or emergent insertion of a temporary transvenous pacing system until a more definitive permanent pacing solution can be established. If patients are hemodynamically stable on presentation, with an underlying stable escape rhythm, chronotropic agents such as isoproterenol or dopamine can be given intravenously until the permanent lead can be repositioned. Twiddler's syndrome can be avoided by minimizing the pocket size, suturing the device to surrounding tissues and, when possible, educating the patient about avoiding external device manipulation. A further preventive measure is using a compression band around the upper chest and shoulder for the first few days after device implantation.^{9,10} Patients at higher risk for twiddler's syndrome (e.g., children, people with intellectual disabilities and people with dementia) should be followed more closely.

Our patient was ultimately brought back to the electrophysiology laboratory, and the lead and generator were repositioned and secured. The patient was discharged home and will be followed by the device clinic.

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For the question, see page 86.

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