

## IMAGE IN CARDIOLOGY

# Twiddler syndrome in a pediatric patient<sup>☆</sup>

## Síndrome de *twiddler* num doente pediátrico

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### Introduction

A seven-year-old girl was diagnosed with transposition of the great arteries with ventricular septal defect and pulmonary stenosis. In the neonatal period, a modified Blalock-Taussig shunt was performed, and at the age of six a Rastelli procedure. Postoperatively she required a permanent VVIR pacemaker (Microny II SR, St. Jude Medical, IsoFlex lead) due to irreversible complete AV block. One year after surgery, she was admitted to the Emergency Department due to pectoral stimulation by the pacemaker. Assessment revealed intermittent ventricular stimulation thresholds (variable and above 2.4V), with unchanged lead impedance. The patient presented advanced AV block. A surgical revision was performed and the pacing lead was extracted by manual traction. There was no macroscopic evidence of lead fracture. A new pacemaker system was inserted (Identity ADx VDR, St. Jude Medical, AV Plus Dx lead), through the right subclavian vein. P and R wave amplitudes were 2 mV and 7.1 mV, respectively, and the ventricular threshold was 0.75 V. The final image showed the pacing lead to be correctly positioned (Figure 1).

Five months later, a further revision revealed inadequate atrial sensing (<0.1 mV), with unchanged ventricular

parameters. On the chest X-ray the pacing lead was stretched and entangled in the pacemaker pocket, which was compatible with twiddler syndrome (Figure 2).

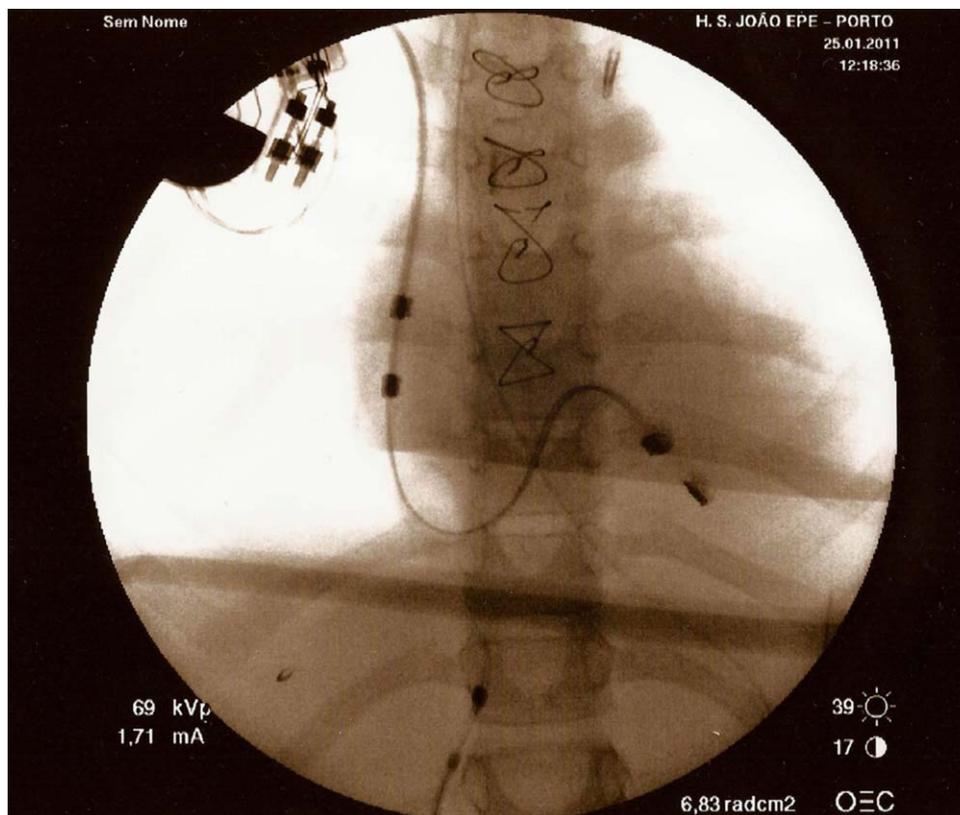
### Discussion

Twiddler syndrome is a rare condition which occurs when a 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 syndrome usually occurs when 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 device, and is seen especially in children and obese and older patients.<sup>1</sup> In the case of a pediatric patient the dislodgement of a pacemaker lead is more problematic because the child’s growth must be taken into account. There is frequently loss of capture and pacemaker malfunction.<sup>2</sup> In many cases it is necessary to explore the pacemaker pocket surgically and replace the lead.<sup>3,4</sup>

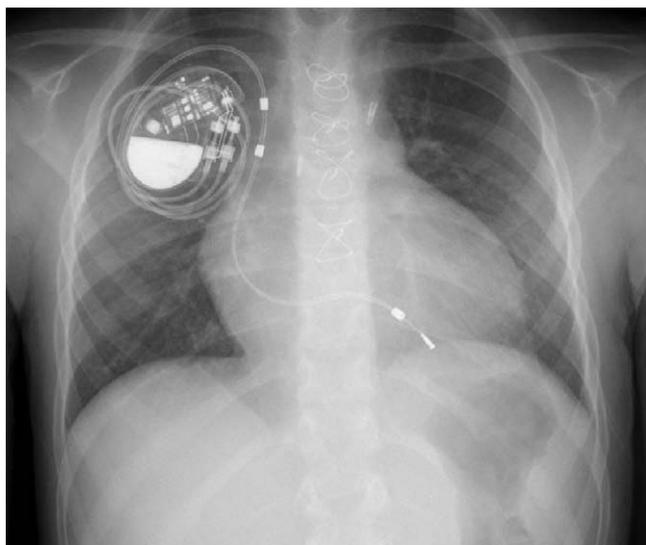
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**Figure 1** Fluoroscopic image following surgical review and pacemaker replacement, with correctly positioned atrial dipole.



**Figure 2** Pacemaker lead without loop in the inferior vena cava and atrial dipole displaced to the superior vena cava, with evidence of "lead twiddling" in the pacemaker pocket.

### Conflicts of interest

The authors have no conflicts of interest to declare.

### References

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