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Twiddler's syndrome in a patient with an implantable-cardioverter defibrillator

A 61 year old patient with an implantable cardioverter-defibrillator (ICD) implanted for ischaemic ventricular tachycardia (VT) was admitted because of sustained VT and several ineffective defibrillator shocks. The ongoing VT was terminated with intravenous procainamide. Multiple episodes of VT were found in the Holter memory of the ICD (upper panel) showing either ineffective shocks or ineffective attempts at antitachycardia pacing (ATP). After each unsuccessful attempt at ATP, electrograms disappeared for variable lengths of time suggesting a contact problem of the electrode. At time of device interrogation lead impedance was high ($> 2000 \Omega$) and neither sensing nor pacing was possible. Fluoroscopy (mid panels) showed coaxial twisting of the lead (red arrow) in the pocket as well as in the heart. Extreme lead twisting was found at surgical exploration (lower panels) with insulation defect and fracture at the proximal segment (black arrow) leading to ineffective stimulation and occasional loss of sensing. The damaged lead was removed without any complication.

Twiddler's syndrome is the result of subconscious, inadvertent or deliberate rotation of a pacemaker or an ICD in its subcutaneous pocket. Potential problems specifically related to ICDs are failure of antitachycardia therapy or inappropriate therapy due to false sensing. Uniquely, our patient developed a stable clinical VT while the lead was partially functioning; it was capable of occasional sensing but failed to deliver effective antitachycardia therapy. Loss of sensing after each ATP attempt was likely the consequence of micromovements caused by local muscle stimulation, indicating the very delicate state of lead integrity.

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