Twiddler's syndrome in children: an unusual cause of pacemaker failure

Simon Abrams, Ian Peart

Figure 1 Case 1. Chest x ray film showing fractured endocardial pacing lead (arrow) (A). Close up of same film showing knot (arrow) behind the pacemaker generator (B). Proximal part of lead with many twists after removal (C).

Abstract

Mechanical causes of pacemaker failure are well recognised. Twiddler's syndrome leading to pacemaker failure has been previously recognised in adults, but there have been no published reports of its occurring in children. Two cases leading to failure of the pacing system are reported. In the first twiddling led to fracture of the lead and in the second it led to displacement of the lead from the heart. Children may be more susceptible to twiddler's syndrome because they have thinner subcutaneous tissues, making leads more accessible, and their comprehension of the consequences may be poor.

(Br Heart J 1995;73:190-192)

Keywords: pacemaker failure, twiddler's syndrome, children.

Mechanical causes of pacemaker failure are well recognised. Fractures of the lead or damage to the insulation can lead to pacemaker malfunction with potentially serious consequences in both children and adults. We report two cases of pacemaker failure due to twiddler's syndrome¹ in children, a previously unrecognised group of twiddlers.

CASE 1

A 5 year old boy was admitted with a short history of intermittent dizziness and syncope. He had been born in 1987 with severe coarctation of the aorta, a bicuspid aortic valve, and a malalignment ventricular septal defect, which were all repaired in infancy. He subsequently developed significant midcavity right ventricular outflow obstruction and severe subaortic stenosis. After surgical relief of the left ventricular outflow obstruction by a septal myectomy, he developed intermittent complete atrioventricular block. A transvenous endocardial pacing system was therefore inserted. A unipolar, tined, steroid tipped lead (Medtronic 4023, Medtronic, Minneapolis, USA) was positioned in the right ventricular apex. At implantation the threshold was 0.3 V at a pulse width of 0.5 ms. A Microminix (Medtronic) VVI unit was attached and implanted in a left prepectoral pouch. Satisfactory pacemaker function was confirmed at regular checks over 18 months.

At this admission he had a heart rate of 100 beats per minute. The pacemaker lead was palpable subcutaneously as a knot in the left deltopectoral groove. An electrocardiogram

Heart Clinic, Royal Liverpool Children's Hospital, Liverpool
S Abrams
I Peart

Correspondence to:
Dr S Abrams, Cardiac
Department, Great Ormond
Street Hospital for Children
NHS Trust, Great Ormond
Street, London WC1 3JH.
Accepted for publication
9 February 1994

Figure 2 Case 2.
Chest x ray film showing
lead in concentric rings
around pacemaker
generator.

showed sinus rhythm with first degree atrio-ventricular block (PR interval 190 ms). There were dissociated pacing artifacts at a rate of 75 per minute. A chest x ray film showed that the pacemaker lead was fractured (figure 1a), and close to the box there was a knot (figure 1b). At the time of replacement the proximal part of the old lead was both fractured and knotted (figure 1c).

CASE 2

A 13 year old girl was referred with twitching of the left arm. She had been born with Turner's syndrome, congenital complete atrioventricular block, and pulmonary stenosis. An epicardial pacing system had been inserted when she was 2 years old and been replaced with an endocardial system when she was 13, one month before presentation. The endocardial system consisted of a tined bipolar ventricular lead (Medtronic 4024) inserted through the left subclavian vein and connected to a Legend II (Medtronic) VVIR pacemaker generator sited in a left prepectoral pouch. Examination showed that her arm was twitching 90 times a minute, corresponding to the programmed rate of the pacemaker. An electrocardiogram confirmed complete atrio-ventricular block. A chest x ray film showed the pacemaker to be surrounded by concentric rings of the pacing lead (figure 2), the entire lead being in the left upper chest. The girl denied any manipulation of either the pacemaker or the lead. Successful reimplantation of the same lead was performed.

Discussion

We report two cases in which the patient's manipulation of a pacing lead was the likely cause of failure of the pacemaker system. In the first case twiddling of the lead led to knotting and fracture; in the second repeated twisting of the generator in a clockwise direction pulled the lead out of the heart and wrapped it around the generator in the left prepectoral pocket. Fortunately, in neither case did this have serious consequences.

Careful examination in both cases gave an indication of the cause. The knot was palpable subcutaneously in the first, and twitching of the arm in the second was suggestive of electrical stimulation of the pectoral muscles.

Pacemaker twiddler's syndrome has been recognised since 1968 with the report of Bayliss *et al.*¹ Several authors have reported similar cases.^{2,3} We have found no reports of the syndrome occurring in children. Lead failure remains an important, if rare cause of pacemaker failure. Till *et al* reported two cases of lead fracture in 24 young children with an endocardial pacemaker system; fracture was due to corrosion in one case and of unknown cause in the second.⁴ Ector *et al* reported six fractured leads in 14 children having multiple systems over 16 years but did not record the causes.⁵

Previous work has shown that helically wound leads are more flexible and resistant to fractures than are straight ones.⁶ The mechanical stresses in a helical pacing lead relates to several factors, and modern leads optimise design features aimed at minimising these stresses. The use of platinum alloys with better shear modules and shear stress characteristics and of smaller diameter wires with increased radius of the helical turns increase lead flexibility.

The site of insertion of the wire into the generator is recognised as a point of critical stress, so the silicon lining in modern leads is thicker at that site. Placing a coil of the lead around the generator also reduces this local stress.

Modern leads do not kink. A determined twiddler, however, will be able to exert stresses that leads are not designed to withstand. A lead that has been in place for some time and is likely to be fixed by fibrous adhesions will be more likely to fracture as a result of twiddling, as in our first case. Leads that have not had time to embed will be more likely to displace from the heart before they fracture, as in our second case.

Children may be more at risk of twiddler's syndrome. Their understanding of the possible consequences of twiddling may be poor and access to the generator and lead is easier because they have thinner subcutaneous tissue. Recognition of the possibility of twiddler's syndrome as a cause of lead fracture or pacemaker failure is important. It is of interest that in most reported cases, patients denied twiddling even when it was witnessed by medical staff.^{4,5} This suggests that it is a subconscious habit. Simple admonition might be insufficient to stop it and, if recurrent, a more detailed psychological approach such as behavioural therapy might be warranted. There were severe social problems in the first case, the behaviour probably being a subconscious habit rather than a deliberate act. In recurrent cases an epicardial system would reduce the child's access to the pacemaker and its lead. The use of a screw in electrode might be worth considering at the time of replacement, although in the second case it was possible to reimplant the original

lead. Another option would be the method advocated by Gillette *et al* of burying the pacing box under the pectoral muscle.⁷ More simple options would be better education of patients and parents and more supervision by parents.

- 1 Bayliss CE, Beanlands DS, Baird RJ. The pacemaker-twiddler's syndrome: a new complication of implantable transvenous pacemakers. *Can Med Assoc J* 1968;99: 371-3.
- 2 Lal RB, Avery RD. Aggressive pacemaker twiddler's syndrome. Dislodgment of an active fixation ventricular pacing electrode. *Chest* 1990;97:756-7.
- 3 Anderson MH, Nathan AW. Ventricular pacing from the atrial channel of a DDD pacemaker: a consequence of pacemaker twiddling? *PACE* 1990;13:1567-70.
- 4 Till JA, Jones S, Rowland E, Shinebourne EA, Ward DE. Endocardial pacing in infants and children 15kg or less in weight: medium-term follow up. *PACE* 1990;13: 1385-92.
- 5 Ector H, Dhooghe G, Daenen W, Stalpaert G, van der Hauwaert L, de Geest H. Pacing in children. *Br Heart J* 1985;53:541-6.
- 6 Thalen HJT, Van der Heide JW, Homan Van der Heide JN, Nieveen J, eds. Transmission of the impulses. In: *The artificial cardiac pacemaker*. 2nd ed. Assen: Royal VanGorcum, 1970;198-210.
- 7 Gillette PC, Edgerton J, Kratz J, Zeigler V. The sub-pectoral pocket: the preferred implant site for pediatric pacemakers. *PACE* 1991;14:1089-92.