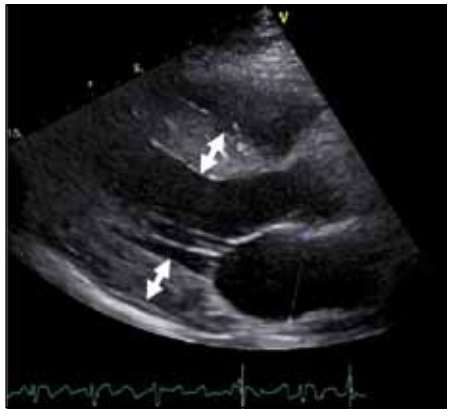

Figure 2. Transthoracic echocardiogram showing a parasternal long axis view with left ventricular hypertrophy (arrows), 1.7 cm in diastole. Ejection fraction 55% and left atrial diameter 3.4 cm.

Santos et al (2010) review the molecular mechanisms causing Friedreich's ataxia.

The net result is a chronic reactive myocarditis with loss of contractile fibres, as opposed to over-expression of contractile fibres as seen in hypertrophic obstructive cardiomyopathy. Coupled with impaired mitochondrial ATP production, this may account for the low voltage electrocardiogram and discrepancies with apparent left ventricular 'hypertrophy' in patients with Friedreich's ataxia.

In terms of presentation this patient, with hypertrophy and an arrhythmia, is not atypical. However, the presence of low voltage in the context of left ventricular hypertrophy on echo, appears unusual. BJHM

Alboliras ET, Shub C, Gomez MR et al (1986) Spectrum of cardiac involvement in Friedreich's ataxia: clinical, electrocardiographic and echocardiographic observations. *Am J Cardiol* **58**(6): 518–24

Child JS, Perloff JK, Bach PM, Wolfe AD, Perlman S, Kark RA (1986) Cardiac involvement in Friedreich's ataxia: a clinical study of 75 patients. *J Am Coll Cardiol* **7**(6): 1370–8

Dutka DP, Donnelly JE, Nihoyannopoulos P, Oakley CM, Nunez DJ (1999) Marked variation in the cardiomyopathy associated with Friedreich's ataxia. *Heart* **81**(2): 141–7

Harding AE, Hewer RL (1983) The heart disease of Friedreich's ataxia: a clinical and electrocardiographic study of 115 patients, with an analysis of serial electrocardiographic changes in 30 cases. *Q J Med* **52**(208): 489–502

Lodi R, Cooper JM, Bradley JL, Manners D, Styles P, Taylor DJ, Schapira AHV (1999) Deficit of in vivo mitochondrial ATP production in patients with Friedreich ataxia. *Proc Nat Acad Sci* **96**(20): 11492–5

Michael S, Petrocine SV, Qian J, Lamarche JB, Knutson MD, Garrick MD, Koeppen AH (2006) Iron and iron-responsive proteins in the cardiomyopathy of Friedreich's ataxia. *Cerebellum* **5**(4): 257–67

Payne RM, Wagner GR (2012) Cardiomyopathy in Friedreich Ataxia: clinical findings and research. *J Child Neurol* **27**(9): 1179–86

Rotig A, De Lonlay P, Chretien D et al (1997) Aconitase and mitochondrial iron–sulphur protein deficiency in Friedreich ataxia. *Nat Genet* **17**(2): 215–17

Santos R, Lefevre S, Sliwa D, Seguin A, Camadro J-M, Lesuisse E (2010) Friedreich ataxia: molecular mechanisms, redox considerations, and therapeutic opportunities. *Antioxid Redox Signal* **13**(5): 651–90

Schadt KA, Friedman LS, Regner SR, Mark GE, Lynch DR, Lin KY (2012) Cross-sectional analysis of electrocardiograms in a large heterogeneous cohort of Friedreich ataxia subjects. *J Child Neurol* **27**(9): 1187–92

LEARNING POINTS

- Friedreich's ataxia is caused by a deficiency of the iron-binding protein frataxin, which results in mitochondrial and nuclear dysfunction.
- The clinical consequences include spino-cerebellar degeneration, diabetes and cardiomyopathy.
- The cardiomyopathy is usually hypertrophic with heart failure a common cause of early death.
- In a proportion of patients cardiomyopathy develops into a dilated cardiomyopathy status with poor systolic function.
- There is currently no cure for Friedreich's ataxia.

Pacemaker in the wrong pocket

This elderly woman (*Figure 1*) informed her daughter that her 'hearing aid' was broken and for weeks she had kept it safe in her trouser pocket. The hearing aid was in fact her VVIR (ventricular sensing, ventricular pacing, inhibited, rate responsive) pacemaker, which had been implanted 6 years previously for complete heart block. The patient was well and haemodynamically stable.

Remarkably, she had no underlying intrinsic cardiac rhythm and was therefore pacemaker dependent. Fortunately, the system was programmed to pace bipolar; as unipolar pacing only works if the generator box is in contact with subcutaneous tissue. Failure to pace would have been fatal in this case. Although skin erosion is common, complete extrusion is a rare phenomenon. BJHM

Dr PD Morris *is Clinical Research Fellow in the Department of Cardiovascular Science, University of Sheffield, Sheffield S10 2JF,* **Dr DZJ Lee** *is Senior House Officer in the Cardiology Department, Northern General Hospital, Sheffield,* **Dr DR Warriner** *is Clinical Research Fellow in the Department of Cardiovascular Science, University of Sheffield, Sheffield, and* **Dr PJ Sheridan** *is Consultant Cardiologist in the Cardiology Department, Northern General Hospital, Sheffield*

Correspondence to: Dr PD Morris (paul.morris@sheffield.ac.uk)

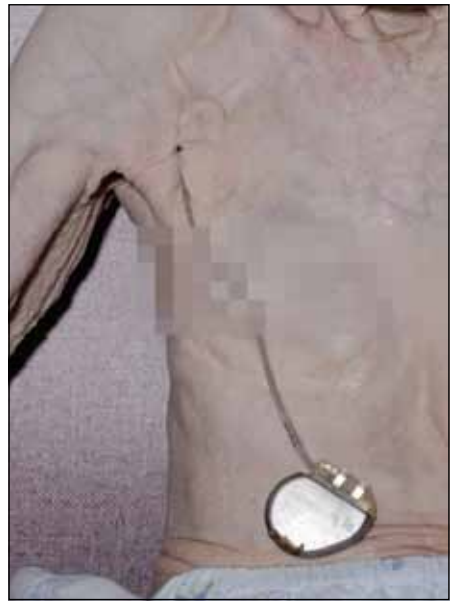

Figure 1. Image showing complete extrusion of the device and wire.