The automatic implantable cardioverter/defibrillator for a life threatening arrhythmia in a case of post-partum cardiomyopathy

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Summary: We report the development of severe life threatening polymorphic ventricular tachycardia in a young woman shortly following her first pregnancy, who ultimately required the insertion of an automatic implantable cardioverter/defibrillator because of the failure of conventional antiarrhythmic therapy. Although only about 7 patients have received units in the UK to date, the experience in the USA, where up to 300 per month may be implanted, suggests that they will become a more common method of treatment in cases of life threatening arrhythmias.

Introduction

The cardiomyopathy associated with pregnancy usually presents in the last month of gestation or in the first 6 months following parturition. The incidence varies geographically from estimates of 1:1,300¹ to 1:4,000² deliveries in the United States to considerably higher in parts of West Africa. Only epidemiological clustering of cardiomyopathy in young women around pregnancy suggests that this is a distinct pathological entity.³ The aetiology remains obscure, and although nutritional, viral, endocrine and immunological mechanisms have been proposed, none has been well supported. Heart failure or embolic phenomena are the commonest presentations, although arrhythmias have been noted.⁴ The mortality of post-partum cardiomyopathy varies in different parts of the world but it has been reported as high as 50% in the USA, the majority of these fatalities occurring in the first 3 months, due to heart failure and systemic pulmonary embolism. Recurrence of the cardiomyopathy is frequent in subsequent pregnancies, which are contraindicated.⁵

Case report

A 34 year old woman presented 5 months after an uncomplicated first pregnancy, when she noticed palpitations at rest and during breast feeding. Over the following month she had 3 episodes of nocturnal

syncope associated with urinary incontinence and subsequent dizziness and fatigue. Clinical examination was unremarkable and she was normotensive, as she had been throughout pregnancy. Initial investigations had sought a cerebral cause, but both resting and nocturnal electroencephalogram, and computed tomographic brain scan, were normal. Cardiovascular investigations were more remarkable; a resting electrocardiogram (ECG) showed widespread T wave abnormalities (Figure 1) and a 24-hour Holter monitor documented episodes of bradycardia-related nonsustained ventricular tachycardia. The 2D echocardiogram revealed a slightly dilated left ventricle and a putative diagnosis of a mild dilated cardiomyopathy was made.

Anti-arrhythmic treatment was commenced with amiodarone which had to be discontinued after 2 weeks because of generalized photosensitivity. Disopyramide controlled her symptoms for 6 months, but thereafter her nocturnal syncopal events recurred and further Holter monitoring demonstrated persistent episodes of ventricular tachycardia. Her therapy was changed to propafenone, which adequately controlled her symptoms for a year. Unfortunately she then had a further syncopal attack and despite the introduction of flecainide she had two daytime attacks, during which time tonic clonic fitting was observed. She was therefore admitted to this hospital for further investigation.

Her resting ECG continued to show widespread ST depression and frequent ventricular ectopics. Her QT interval was normal. Short runs of ventricular tachycardia were observed. An exercise test failed to induce any arrhythmias and indeed abolished resting

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Figure 1 Resting electrocardiogram showing the widespread ST abnormalities.

ventricular extrasystoles, but produced 4 mm ST segment depression in the inferior and anteroseptal leads, which occurred in the absence of chest pain. Further ambulatory monitoring revealed frequent polymorphic ventricular tachycardia, the morphology of which suggested torsade de pointes. The 2D echocardiogram now only showed minimal ventricular dilation and the MUGA left ventricular ejection fraction was 41% (RV ejection – 21%). Left ventriculography and coronary arteriograms were normal and she had normal right and left sided pressures. Electrophysiological studies, performed under general anaesthetic, failed to induce sustained ventricular tachycardia despite aggressive stimulation protocols before and after isoprenaline. Investigations for autoimmune, endocrine, metabolic and viral aetiologies were negative.

During hospital admission, when antiarrhythmic medications were stopped, she had nine further episodes of documented ventricular tachycardia producing syncope. All but three episodes were terminated spontaneously or with a few minutes of external cardiac massage. On three occasions ventricular tachycardia degenerated into ventricular fibrillation and DC cardioversion was required (150J). Attacks persisted despite therapy with flecainide, nadolol and disopyramide. A clear observation was made between bradycardia and the inception of the ventricular tachycardia and this was exacerbated by beta-blockade. In view of this a temporary atrial pacing wire was inserted and set at 90 beats per minute, following which no further syncopal episodes occurred.

In view of the failure of antiarrhythmic drugs to control her ventricular tachycardia and the absence of a focus of ectopic activity which would be suitable for either transvenous or surgical ablation, it was decided to insert an automatic implantable cardioverter/defibrillator (AICD). Under general anaesthetic, a bipolar endocardial lead was positioned in the right ventricle, and two ventricular patch leads were sewn over the apex (anode) and the posterior lateral wall (cathode) of the left ventricle, external to the fibrous pericardium (which was not opened) via a left thoracotomy. The pulse generator was located in a pocket in the upper abdominal wall behind the rectus sheath. Following insertion of the AICD, slow release disopyramide was commenced and, to date, there have been no further episodes of syncope. Holter monitoring showed ventricular extrasystoles but no episodes of ventricular tachycardia. The mean heart rate on the disopyramide remained above 70 beats per minute, even during the night. One week following the insertion of the device testing was performed under general anaesthetic. Ventricular fibrillation was induced and the device successfully sensed the arrhythmia and cardioverted the patient with the first shock. The AICD has not discharged spontaneously in 12 weeks of follow-up.

Discussion

Mechanism of action of AICD

Automatic implantable cardioverter/defibrillators recognize ventricular arrhythmias by sensing heart

rate with or without ECG morphology. Electrical countershock is then delivered, when appropriate, through special electrodes. Bipolar endocardial or myocardial leads sense R waves and determine when the rate exceeds a preset level for the device. Morphological determination (which is optional) examines the percentage of the time the ECG is isoelectric via ventricular patch leads, which are also used for defibrillation. During ventricular fibrillation continuous electrical activity prevents the ECG signal from returning to the baseline – the absence of a significantly long isoelectric period can be used to detect ventricular fibrillation. The AICD requires 5–20 seconds to diagnose ventricular tachyarrhythmias, and a further 5–15 seconds for the pulse generator to charge its storage capacitors. The AICD will then deliver a truncated exponential pulse of 3–8 ms duration and an output of approximately 700 volts. This delivers 23–28 joules across the epicardial ventricular patch leads. The time elapsed between the onset of an arrhythmia and initial countershock is 10–35 seconds. If the recognized arrhythmia persists, 3 further shocks of 28–37 joules will be delivered after the same sequence of detection and charging has been accomplished. After 4 discharges, the AICD will not pulse again unless there is an intervening 35 seconds of a different rhythm to which the device does not respond. This rhythm may be sinus rhythm, heart block, bradycardia, idioventricular rhythm or asystole. Thereafter the 4 shock sequence will recommence. The functional status of the unit, and the number of shocks it has delivered may be analysed noninvasively by the use of an external magnet and radio frequency telemetry.

Use and problems of AICD

AICD devices are intended to prevent sudden cardiac death due to ventricular tachyarrhythmias. Suitable patients include survivors of sudden death due to ventricular arrhythmias that have occurred outside the context of acute myocardial infarction and patients with recurrent haemodynamically significant ventricular tachycardia resistant to drug therapy and/or surgical or electrical ablation.

These units are not without drawbacks. Implantation site infections and lead migration have been described as in other pacemaker systems. Inappropriate detection may occur particularly in the presence of rapid supraventricular tachycardias which may trigger the unit.⁶ Similar problems may arise in the presence of implanted demand pacemakers because the AICD may sense the pacemaker stimulus.⁷ Theoretical problems with electromagnetic interference may also trigger the unit, or switch it to its inactive mode, and so diathermy and magnetic resonance imaging are contraindicated. The unit may

discharge whilst the patient is conscious causing considerable discomfort. This may occur due to faulty sensing of supraventricular tachycardia, interactions with pacemakers or with self-terminating ventricular tachycardias. If ventricular tachycardia lasts 5–20 seconds the unit will diagnose the arrhythmia and then takes 5–15 seconds whilst charging, before delivering the shock. Thus spontaneously self-terminating ventricular tachycardia of duration 5–35 seconds may theoretically trigger the unit, revert to normal rhythm and then the unit will discharge.

Persons administering cardiopulmonary resuscitation are reported to have been startled by skin surface potentials from patients during discharge of the AICD units and there is a theoretical risk of inducing arrhythmias in the resuscitator. The presence of epicardial defibrillating leads may insulate the heart from standard external defibrillation, and in these circumstances external defibrillation should be delivered perpendicular to the line of the ventricular patch electrodes.

The AICD appears to be of the greatest value when used in conjunction with other antiarrhythmic therapies which decrease, but do not abolish, the incidence of tachycardia in the patient. In this case the pericardium was not opened so as not to prejudice future attempts at curative surgery. Sole usage of the device in patients with frequent ventricular tachycardia will cause rapid battery depletion. The manufacturers indicate that the units have a lifelong output capacity of 100 shocks of 25J and recommend unit replacement at least every 2 years, at a cost currently of £12,000. This cost however seems to be justified since there is an important improvement in survival.

Echt⁷ implanted 70 AICD devices in patients with ventricular arrhythmias resistant to antiarrhythmic drugs who experienced cardiac arrest or recurrent life threatening ventricular tachycardia. One year cardiac death rate was 10.1% and sudden death rate was 1.8% which compares favourably with historical controls from the same institution where the 1 year mortality in patients with ventricular tachyarrhythmias resistant to drugs and electrosurgery was 40% for cardiac death and 26% for sudden death.⁸

When the early AICD was introduced by Mirowski, only a single pair of electrodes were used to sense the arrhythmias on morphological criteria alone and deliver a nonsynchronised shock.⁹ These units detected ventricular fibrillation and rapid ventricular tachycardia only. Subsequent design modifications including the use of a second pair of electrodes for rate sensing, and synchronization of cardioversion has now allowed successful treatment of slower ventricular tachycardias also. Future modifications are likely and will include the addition of a memory function which allows identification of the triggering

stimuli which cannot be currently documented. This is important since subsequent adjustment of the sensing parameters can then be made if the stimulus causing the device to discharge is not a ventricular tachyarrhythmia, but a supraventricular arrhythmia and adequate differentiation may then be made possible by

manipulation of the device, by the provision of individually programmable sensing windows. The incorporation of antibradycardia pacing into these units may also increase their clinical value and refining the lithium batteries to extend life would reduce the need for unit replacement.

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