A 41 year old woman with type 2 diabetes, hypertension, and hyperlipidaemia but no known heart disease received 130 DC shocks for repeated cardiac arrests due to ventricular tachyarrhythmias over 48 hours. She was stabilised by intravenous amiodarone and had a defibrillator implanted. Serial ECGs did not change, but raised troponin I confirmed myocardial infarction as the underlying cause. Electrical storm is an uncommon and dramatic but usually treatable syndrome of recurrent ventricular arrhythmias. Frequent precipitants of electrical storm include recent worsening heart failure, hypokalaemia, hypomagnesaemia and myocardial ischaemia. Amiodarone is the antiarrhythmic agent of choice and implantable cardioverter defibrillator improves long term outcome.

Electrical storm is an uncommon and dramatic but usually treatable syndrome of recurrent ventricular arrhythmias. These critically ill patients require care directed at different goals. They need immediate resuscitative treatment, identification of the underlying cause and strategies for long term prevention of recurrence. We present a case of electrical storm when a woman was defibrillated 130 times over 48 hours but went on to make a good recovery.

CASE REPORT

A 41 year old woman with type 2 diabetes, hypertension, hyperlipidaemia, obesity, and diabetic nephropathy was admitted for assessment of breathlessness. There was no history of ischaemic heart disease. Initial investigations showed haemoglobin 7.4 g/dl, urea 20.5 mmol/l, and creatinine 291 mmol/l, and normal electrolytes. Chest radiography revealed pulmonary oedema. ECG showed sinus rhythm with poor progression of R waves in V1-V4. QT interval was normal (fig 1). She received blood transfusion and diuretics for heart failure and was symptomatically improving.

A week after admission, she had an unexpected asystolic cardiac arrest, which reverted with cardiopulmonary resuscitation. Pulmonary embolism was considered. While spiral computed tomography was being done, she had recurrent pulseless ventricular tachycardia (VT) (fig 2) and ventricular fibrillation (VF). Cardiopulmonary resuscitation was started and she received more than 40 DC shocks before achieving sinus rhythm with a cardiac output. She was started on intravenous amiodarone. The spiral computed tomogram was negative.
She continued to have repeated episodes of pulseless VT and VF requiring defibrillations. Serum potassium and magnesium were normal and arterial blood gas pressures showed hypoxia and metabolic acidosis. She became hypotensive and oliguric. Inotropic support with adrenaline (epinephrine) and dobutamine was started and she was ventilated. She remained unstable and had multiple episodes of ventricular arrhythmia, cardiac arrests, and defibrillations over the next 24 hours. Though serial ECGs remained unchanged, troponin I was raised at 12.94 μg/l (reference range 0–0.1) confirming myocardial infarction.

Overdrive pacing was withheld as the arrhythmia settled over the next 24 hours, she was extubated and her urine output improved. She had received 130 DC shocks over 48 hours. Echocardiogram showed impaired left ventricular function with apical akinesis. Coronary angiography showed triple vessel disease unsuitable for revascularisation. Ejection fraction was 30%. A defibrillator was implanted and oral amiodarone was continued. At follow up three months later, she was in sinus rhythm and the defibrillator had not discharged. She was taking spironolactone and carvedilol for left ventricular dysfunction. ACE inhibitors were withheld in view of renal impairment.

DISCUSSION

Electrical storm is defined as recurrent, haemodynamically destabilising ventricular tachycardia or ventricular fibrillation occurring two or more times over a 24 hour period, and usually requiring electrical cardioversion or defibrillation. It occurs most often in patients with coronary artery disease, including those with prior myocardial infarction, left ventricular dysfunction, and ventricular dilatation. Frequent precipitants of electrical storm include recent worsening heart failure, hypokalaemia, hypomagnesaemia, and myocardial ischaemia. Silent myocardial ischaemia is known to be common in diabetes and carries the same risk and prognosis as symptomatic coronary artery disease. This patient did not have chest pain and serial ECGs did not show evolving changes. Myocardial infarction, confirmed by the rise in troponin I, was the precipitating cause of electrical storm. Cardiac troponin I is a sensitive and specific marker of myocardial injury. Myocardial infarction and values are normal, or only minimally raised, after repeated cardioversions. Defibrillation is a priority if the patient is in cardiac arrest with pulseless VT or VF but attempts must also be made to find underlying causes and prevent further episodes. Treatment of electrical storm with conventional antiarrhythmic drugs is seldom effective. Amiodarone, an antiarrhythmic agent with established efficacy in the treatment of a variety of atrial and ventricular tachyarrhythmias, has been used in this setting. Three recent studies examined the effect of intravenous amiodarone in the management of electrical storm. In a study of critically ill patients with coronary artery disease, intravenous amiodarone was effective in controlling life threatening ventricular tachyarrhythmias in 50% patients in first 24 hours and 64% patients in the second 24 hours. A large, randomised trial found amiodarone superior to bretylium in patients with electrical storm. Overall survival was 86% at 48 hours, substantially better than would be expected from historical controls.

In sustained VT or VF, the implantable cardioverter defibrillator (ICD) is highly effective in treating recurrences. ICD therapy improves survival in patients who have survived cardiac arrest and are at risk of sudden death—that is, with a history of myocardial infarction and reduced systolic left ventricular function. In the Multicentre Automatic Defibrillator Implantation Trial (MADIT), in patients with myocardial infarction at high risk of ventricular arrhythmias, prophylactic therapy with an ICD improved survival compared with conventional medical treatment (hazard ratio for mortality 0.46, p=0.009). There was no evidence that amiodarone, β blockers, or any other anti-arrhythmic therapy had a significant influence on the observed hazard ratio. The Antiarrhythmics versus Implantable Defibrillators (AVID) investigators found overall survival at three years with ICD to be 75% compared with 64% on medical treatment, predominantly amiodarone, in patients resuscitated from ventricular arrhythmias. ICD therapy has been shown to reduce all cause mortality by 23% as compared with amiodarone/metoprolol in survivors of cardiac arrest secondary to recorded ventricular arrhythmias. A meta-analysis of ICD in secondary prevention in survivors of ventricular fibrillation/sustained ventricular tachycardia showed 28% reduction in the relative risk of death as compared with amiodarone. This is almost entirely attributable to a 50% reduction in arrhythmic death. Patients with left ventricular ejection fraction less than or equal to 35% derive significantly more benefit from ICD therapy.

Antiarrhythmic drug therapy in patients with ICDs has decreased over the past decade. There have not been any controlled clinical trials on the role of antiarrhythmic drugs after implantation of defibrillators. In addition to prevention of VT, drug therapy may slow tachycardia rate and make it more tolerated haemodynamically. Some of the class III antiarrhythmic drugs may actually lower defibrillation threshold. Unfortunately, these drugs may also have adverse interactions with ICDs. An increase in defibrillation threshold, proarrhythmic effects, and bradycardic effects may be undesirable. Detailed electrophysiological evaluation after drug treatment is started is recommended for most patients with ICD.

This case illustrates the importance of defibrillation and amiodarone in management of electrical storm. Despite repeated defibrillations, she went on to make a good recovery. The outcome of such patients may not be as grim as previously supposed and aggressive supportive treatment is indicated. With newer approaches and treatments, there may indeed be sunshine after the storm.

Contributors
P K Moulik, E L Rose, and A A Khaleeli were involved in clinical care of the patient. P K Moulik and M N Attar wrote the paper with supervision of A A Khaleeli and E L Rose. A A Khaleeli will act as the guarantor of the paper.

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REFERENCES


