ELECTRICAL STORMS AND THEIR PROGNOSTIC IMPLICATIONS

Zahid Aslam Awan, Mahmood ul Hassan, Kamran Bangash, Bakhtawar Shah, Lubna Noor

Department of Cardiology, Postgraduate Medical Institute, Hayatabad Medical Complex, Peshawar, Pakistan

Background: Prevention of sudden cardiac death has always been a challenge for electrophysiologists and to date, automatic implantable cardiovertor defibrillator (AICD) is found to be the only remedy. This device delivers an intracardiac shock whenever it senses a fatal ventricular arrhythmia in order to achieve sinus rhythm. If the delivery of these intracardiac shocks becomes frequent, the situation is declared as an electrical storm. This article deals with the frequency, precipitating factors and prevention of electrical storms. **Methods:** One hundred and ten episodes of electrical storms (a total of 668 shocks) were retrospectively analysed in 25 recipients of automatic implantable cardioverter defibrillators. ECG, echocardiography, serum electrolytes, urea and creatinine were done for all the patients, and they were hospitalized for a minimum of 24 hours. **Results:** During the 3 year study period, all the 25 patients with an implantable cardiovertor defibrillator, on an average, received one shock per two years. However, 12 out of these 25 patients (50%) had more than two shocks within 24 hours. Most of these patients with electrical storms were having active ischemia, electrolytes imbalances or renal failure. **Conclusion:** Electrical storms are common in patients with coronary artery disease with impaired left ventricular functions. Ischemia, electrolytes imbalances and renal failure predispose to the electrical storms. Electrical Storms are predictors of poor prognosis.

Keywords: Electrical Storm (ES), Automatic Implantable Cardiovertor Defibrillator (AICD), Ventricular Tachycardia (VT), Ventricular Fibrillation (VF)

INTRODUCTION

Coronary artery disease (CAD) is the leading cause of human mortality all over the world and sudden cardiac death (SCD) is the most common presentation of CAD. Numerous drugs have been tried and till now the consensus is that antiarrhythmic drugs are either harmful or have no effect in SCD.[1,2]

The automatic implantable cardioverter defibrillator (AICD) is the only invention of the twentieth century which has revolutionized the treatment of patients at risk for sudden cardiac death.[3–6] Initially introduced in humans in 1980,[7] and approved by the Food and Drug Administration (FDA) in 1985, the ICD has evolved from a treatment of last resort to a first-line treatment and prophylactic therapy for patients at risk for ventricular tachycardia (VT) or ventricular fibrillation (VF).

Indications for AICD implantation can be divided into 2 broad categories: secondary prophylaxis against sudden cardiac death, i.e., as initial therapy in survivors of cardiac arrest due to ventricular fibrillation or hemodynamically unstable ventricular tachycardia, and primary prophylaxis in patients with CAD having ejection fraction of less than 35%, cardiomyopathies with ejection fraction less than 35%, hypertrophic obstructive cardiomyopathy, Brugada syndrome, Long QT syndrome and arrhythmogenic right ventricular cardiomyopathy.

Approximately 50% to 70% of patients treated with an AICD receive appropriate device-based therapy within the first 2 years following implantation. Most arrhythmic events require only one appropriate ICD firing for termination.[7] However, some patients receive multiple appropriate shocks during a short period of time. This condition has been referred to as an 'electrical storm' (ES), which has been defined as recurrent VT/VF occurring two or more times in a 24-hour period, and usually requiring device intervention in the form of electrical cardioversion/defibrillation or antitachycardia pacing.[8] ES occurs in approximately 25% of ICD patients within 3 years, with typically 5–55 individual VTs within one storm. Potential triggers can be found in approximately 66% of patients and include new/worsened heart failure, changes in antiarrhythmic medication, context with other illness, psychological stress, diarrhoea, and hypokalemia.[9,10]

One of the risks of ICD implant is that of inappropriate ICD shocks, defined as one that is not precipitated by accurate detection of a malignant ventricular arrhythmia, VT, or VF. Typically, inappropriate ICD shocks result when atrial arrhythmias(such as atrial fibrillation, atrial tachycardia, or atrial flutter) accelerate the ventricular rate beyond the set limit for delivery of ICD shock therapy, and may also result from sinus tachycardia, supraventricular tachycardia (SVT), substance abuse, electromagnetic interference, and ventricular oversensing.[10,11] Occasionally, however, recipients of ICD perceive false impression of getting intracardiac shocks, which after device interrogation, is revealed that there has been no arrhythmic event, and hence no shock at all. This phenomenon is referred to as 'phantom shocks'.[5]

The precise time of occurrence of ES after AICD implantation, appropriate clinical management of these patients, and prognostic implications of electrical storm are yet ill defined. Thus, the present retrospective observational study is aimed at answering these questions as much as possible in a cohort of 25

consecutive patients who had received contemporary AICDs, and presented with an electrical storm at our institution (which is the only centre in the region to offer AICD implants and has maintained a registry for proper follow-up).

MATERIAL AND METHODS

From January 2006 to December 2009, whenever, any of the recipients of AICDs was received in the emergency room with one or more shocks, they were immediately admitted into Coronary Care Unit. The patient was considered for the present analysis, and device interrogation was carried out within 24 hours of the event.

- VF was defined as an event with a ventricular rate of more than 188 bpm.
- VT was defined as an episode with a ventricular rate of more than 150 bpm.
- ES was defined for the purpose of this analysis as the occurrence of ventricular tachycardia or fibrillation resulting in device intervention (antitachycardia pacing and/or shock delivery) three or more times within a 24-hours period.
- For each arrhythmia episode, the appropriateness of ICD therapy was also verified by device interrogation.

Patients were retained in CCU under strict vigilance for a minimum of 24 hours period. An ECG and echocardiogram was recorded, and serum electrolytes, urea, creatinine and blood sugar were sent for. If any underlying cause was revealed, it was immediately corrected. If despite this, the shocks would persist, medical therapy with infusion amiodarone (150 mg stat over 10–15 minutes, then 360 mg over the next 6 hours and the 540 mg over the next 12–18 hours) was commenced. Any other adjuvant therapy, as per guidelines and recommendations, was administered as and when needed, such as beta-blocker therapy, isoproterenol, magnesium, or over-drive pacing.

RESULTS

Table-1 shows the basic clinical characteristics of the 25 recipients of AICD who presented to our centre with electrical storms. Their mean age was 52±17 years, 22 of them were male and 3 female. Three of these AICDs were dual-chambered, while the rest of the 22 were single-chambered. The minimum number of days for the first device therapy after implantation was 200. All of the patients, on an average, received one shock per two years. However, 12 out of these 25 patients (50%) had more than two shocks within 24 hours. Careful analysis of AICD stored electrograms revealed that patients with electrical storms were more likely to have had VF their index arrhythmia (80% VF as compared to 20% VT). Hypokalemia due to diarrhoea was the triggering factor in 2 patients, hyperkalemia secondary to renal failure in 4 patients, 14 patients were having active ischemia induced VT/VF, while no precipitating cause was found in 5 patients. No episode required external resuscitation. Shock alone was delivered to treat all episodes of VF, while for VT, antitachycardia pacing alone or in combination with shock was employed by the device. When admitted into CCU, infusion amiodarone was given to all patients, and 100% response was seen to it. Two events of ES were found to be phantom shocks.

Table-1: Basic clinical characteristics of the 25 patients presented with ICD shocks/ Electrical Storms at our centre

Total number of patients	25
Total number of ES* received	110
Total number of shocks delivered	668
Number of patients who had ES*	13 (52%)
Number of patients who had shocks but not ES*	10 (40%)
Number of patients with inappropriate shocks	0
Number of patients with phantom shocks	2 (8%)
Presenting arrhythmia/s	
VF**	535 (80%)
VT***	133 (20%)
Gender	
Male	22 (88%)
Female	3 (12%)
Underlying heart disease	
Coronary artery disease	16 (64%)
Congestive cardiomyopathy	3 (12%)
Brugada syndrome	3 (12%)
Long QT syndrome	1 (04%)
Ebstein's anomaly	1 (04%)
Arryhthmogenic right ventricular cardiomyopathy	1 (04%)
Precipitating cause for ES*	
Active ischemia	14 (56%)
Hyperkalemia	4 (16%)
Hypokalemia	2 (08%)
LQTS	0
Unknown	5 (20%)
Response to amiodarone	**100%**

*Electrical storm, **Ventricular fibrillation, ***Ventricular tachycardia

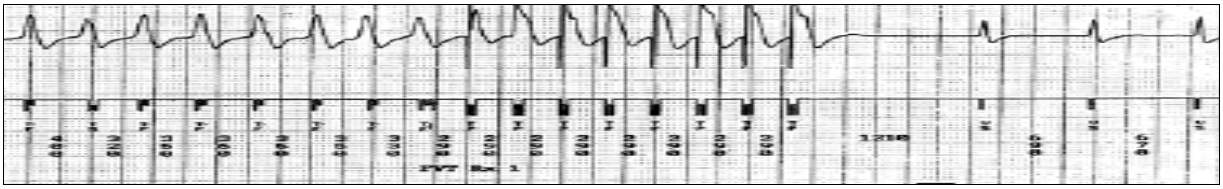

Figure-1: ICD recording during ventricular tachycardia. Far-field electrogram recording (top), marker channel depicting device interpretation (middle), and cycle length of each complex (bottom).

The episode of ventricular tachycardia (labelled TF) is detected and antitachycardia pacing (TP) is delivered, terminating the arrhythmia.

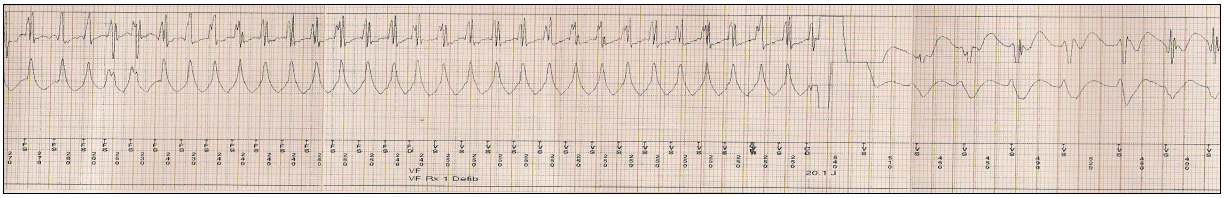

Figure-2: VF being detected at a rate of 200 bpm which has been terminated and reverted to sinus rhythm after an intracardiac shock of 20J is delivered

DISCUSSION

CAD is the leading cause of human morbidity and mortality all over the world, and most of the SCDs[7] are caused by ischemia induced ventricular arrhythmias[1]. Current consensus is that device therapy (AICD) is more effective in preventing SCDs,[8,10] especially in patients with impaired LV functions. As there is a growing epidemic of CAD, therefore, the recommendations for AICD implantation are accordingly becoming stronger.[11]

The incidence of electrical shocks is 50% in our patients which is much higher than that reported[12] in the literature. The most important cause for it is that all of our patients received AICD for secondary prevention of SCD as compared to western countries where AICD is employed for primary prevention mainly.[8,9] Hence, ES is a rarer event in such recipients of AICD for primary prevention of SCD.[12]

Another contributing factor is the relative lack of or delay in provision of coronary revascularization/ reperfusion therapy, due to poor economical conditions and lack of awareness. These results in poor left ventricular functions, providing a more favourable ventricular substrate for fatal arrhythmias, hence, resulting in a higher incidence of shocks delivered by the devices. Also, patients with such hypoperfused myocardial substrate are unable to combat with stress, and even minimal amount of stress such as exercise can provoke ischemia-induced ventricular arrhythmias resulting in frequent device based therapy.

Inadequate use of beta blockers and statins in our patients with ischemic cardiomyopathy and AICD, is another contributing factor for a higher incidence of device therapy. Beta blockers also play a protective role against occurrence of ventricular arrhythmias.[13] Drug therapy with statins, has been shown to reduce, by more than half, the frequency of inappropriate ICD shocks secondary to occurrence of atrial fibrillation and atrial flutter.[14] Electrolytes imbalances due to drugs like diuretics and diseases like diarrhoea and renal impairment, are also commoner in our population, which again contributes to occurrence of ventricular arrhythmias, and initiating device-based shocks and electrical storms.

It has also been in our study that there was no incidence of inappropriate shocks in our patients. It can be explained on the basis that almost all of our patients are on amiodarone therapy, thereby, suppressing the supraventricular arrhythmias.[15] It is now advocated that patients with frequent occurrence of ES despite medical therapy, should be subjected to substrate mapping and radiofrequency ablation of the VT.[16,17] Nowadays, even heart transplantation has been proposed for such AICD recipients.[18]

Electrical storms always underlie a change in the ventricular substrate by various factors, which result in a sudden increase in arrhythmia incidence deviating from background sporadic arrhythmic events and causing multiple intracardiac shocks. Such arrhythmias and recurrent ICD interventions are responsible not only for further impairing the myocardium, but also drains the AICD battery. An ICD shock is generally very painful, and such recurrent shocks can render dramatic psychological repercussions on the patient.[10] As recurrent ES are responsible for an important morbidity, there is a possible role for prophylactic interventions. An earlier revascularization procedure should be carried out in order to reduce ischemia induced arrhythmic events. Optimal adjuvant drug therapy (amiodarone, beta blockers and statins) should be given to these patients in order to reduce the frequency of ESs. Patients with ICDs should be offered emotional or psychological support for anxiety, depression, and difficulties in adjusting to life with an ICD. The recipients of AICD should be observed in a dedicated ICD clinic. Studying patterns of arrhythmia recurrence may be valuable; characteristics of repeated VT/VF episodes may lead to a better understanding of factors responsible for arrhythmia induction, and may identify a subpopulation at high risk. Systematic programming of AICD may avoid unnecessary shocks (long VT detection, antitachycardia pacing where ever possible) which otherwise can fuel the sympathetic tone and prolong ES.[19] Therapy of repetitive VT/VF may include reversing the precipitating cause, anti-arrhythmic drugs, and catheter ablation, which will very clearly reduce the burden of ES. If individuals at risk can be identified, prophylactic interventions might be developed which will improve the quality of patient's life.

CONCLUSION

Electrical storms are a commoner event in CAD, especially in ongoing ischemia and with impaired left ventricular functions. There are identifiable precipitating

causes for ESs which should always be looked for, prevented and corrected as early as possible. It is evident that life expectancy of patients with AICD who suffer frequent ESs is reduced. ES, therefore, implicates a bad prognosis in recipients of AICD. Drug therapy with amiodarone is very effective.

REFERENCES

1. Greene HL, Roden DM, Katz RJ, Woosley RL, Salerno DM, Henthorn RW. The Cardiac Arrhythmia Suppression Trial (CAST). J Am Coll Cardiol 1992;19:894–8.
2. Mark DB, Nelson CL, Anstrom KL, Al khatib SM, Tsiatis AA, Cowper PA. Cost effectiveness of defibrillation therapy or amiodarone in chronic stable heart failure: Results from the Sudden Cardiac death in Heart Failure Trial (SCD-HeFT). Circulation 2006;114(2):135–42.
3. Antiarrhythmics versus Implantable Defibrillators (AVID) Investigators. A comparison of antiarrhythmic-drug therapy with implantable defibrillators in patients resuscitated from near-fatal ventricular arrhythmias. N Engl J Med 1997;337:1576–83.
4. Connolly SJ, Gent M, Roberts RS. Canadian Implantable Defibrillator Study (CIDS): a randomized trial of the implantable cardioverter defibrillator against amiodarone. Circulation 2000;101:1297–302.
5. Moss AJ, Hall WJ, Cannom DS. Improved survival with an implanted defibrillator in patients with coronary disease at high risk for ventricular arrhythmia: Multicenter Automatic Defibrillator Implantation Trial Investigators. N Engl J Med 1996;335:1933–40.
6. Buxton AE, Lee KL, Fisher JD. A randomized study of the prevention of sudden death in patients with coronary artery disease: Multicenter Unsustained Tachycardia Trial Investigators N Engl J Med 1999;341:1882–90.
7. Mirowski M, Reid PR, Mower MM. Termination of malignant ventricular arrhythmias with an implanted automatic defibrillator in human beings. N Engl J Med 1980;303**:**322–4.
8. Villacastin J, Almendral J, Arenal A. Incidence and clinical significance of multiple consecutive, appropriate, high-energy discharges in patients with implanted cardioverter defibrillators. Circulation. 1996;93:753–62.
9. Israel CW, Barold SS. Electrical storm in patients with an implanted defibrillator: matter of definition. Ann Noninvasive Electrocardiol 2007;12(4):375–82.
10. Credner SC, Klingenheben T, Mauss O, Susanne C. Electrical storm in patients with transvenous implantable cardioverter-defibrillators: incidence, management and prognostic implications. J Am Coll Cardiol 1998;32:1909–15.
11. Derek V. Exner, Sergio L. Pinski, D. Electrical Storm Presages Nonsudden Death the Antiarrhythmics versus Implantable Defibrillators (AVID) Trial. Circulation 2001;103;2066–71.
12. Moss AJ, ZarebaW, Hall WJ, Klein H, Wilber DJ, Cannom DS. Prophylactic implantation of a defibrillator in patients with myocardialinfarction and reduced ejection fraction. N Engl J Med 2002;346:877–83.
13. Kendall MJ, Lynch KP, Hjalmarson, Kjekshus J. Beta-blockers and Sudden Cardiac Death. Ann Intern Med 1995;123:358–67.
14. Vyas AK, Guo H, Moss AJ, Olshansky B. Reduction in ventricular tachyarrhythmias with statins in the Multicenter Automatic Defibrillator Implantation Trial (MADIT)-II. J Am Coll Cardiol 2006;47(4):769–73.
15. Herre JM, Sauve MJ, Malone P. Long term results of amiodarone therapy in patients with recurrent sustained ventricular tachycardiaor ventricular fibrillation. J Am Coll Cardiol 1989;13:442–9.
16. Avitall B, Port S, Gal R. Automatic implantable cardioverter/ defibrillator discharges and acute myocardial injury. Circulation 1990;81:1482–7.
17. Carbucicchio C, Santamaria M, Trevisi N, Maccabelli G, Giraldi F, Fassini G. Catheter Ablation for the treatment of Electrical Storm in Patients with Implantable Cardioverter-Defibrillators. Circulation 2008;117:462–9.
18. Barnay C; Taieb J; Morice R. Electrical storm. Ann Cardiol Angeiol (Paris) 2007;56(5):183–7.
19. Israel CW; Barold SS. Electrical storm in patients with an implanted defibrillator: a matter of definition. Ann Noninvasive Electrocardiol 2007;12(4):375–82.

Address for Correspondence:

Dr. Zahid Aslam Awan, Associate Professor of Cardiology, Hayatabad Medical Complex, PGMI, Peshawar, Pakistan. **Cell:** +92-300-5940698

Email: zahidccf@hotmail.com