

# ELECTRICAL STORMS IN PATIENTS WITH IMPLANTABLE CARDIOVERTER DEFIBRILLATOR

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**Abstract:** *Electrical storm is defined as the occurrence of three or more distinct episodes of ventricular tachycardia or ventricular fibrillation in 24 hours, requiring the intervention of the defibrillator. The aim of the paper is to present a case-series of patients with electrical storm requiring hospitalization. The patients were collected from a retrospective study of 217 patients with ICD implanted at ICCO clinics between 01.02.2009-30.04.2014. We analysed retrospectively the patient's files and the recordings from ambulatory follow-up visits.*

**Key words:** *electrical storm, implantable cardioverter-defibrillator, ventricular tachycardia, ventricular fibrillation.*

## 1. Introduction

Implantable defibrillators are lifesaving and have improved mortality rates in patients at risk of sudden death, both in primary and secondary prevention. However, these devices are unable to modify the myocardial substrate, which remains susceptible to life-threatening ventricular arrhythmias.

Electrical storm (ES) is defined as the occurrence of three or more distinct episodes of ventricular tachycardia or ventricular fibrillation in 24 hours, requiring the intervention of the defibrillator [10].

## 2. Aim

To evaluate the characteristics, clinical presentation and outcome of patients with electrical storm requiring hospitalization.

## 3. Materials and Methods

We assessed the prevalence, features, and predictors of ES in 217 patients with ICD implanted at ICCO clinics between 01.02.2009-30.04.2014. We analysed retrospectively the patient's files and the recordings from ambulatory follow-up visits. Each patient was followed-up one month after the implant and every three months thereafter. The programmed parameters were every time analysed and changed if needed. At each interrogation we also evaluated the number and type of arrhythmia – related events (sustained/non-sustained ventricular tachycardia, number of anti-tachycardia pacing therapies (ATP) delivered, number and appropriateness of shocks. We also have collected data regarding the re-hospitalization. If the patient was missing from follow-up visit,

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we made calls to find out the patient status (hospitalized or death).

Implant indication was made according to guidelines. Resynchronization therapy (CRT-P) or resynchronization therapy with defibrillator support (CRT-D) was recommended for reducing the morbidity and mortality in patients with heart failure NYHA III or ambulatory IV who had an left ventricular ejection fraction (LVEF)  $\leq$  35% and QRS  $\geq$  120 msec and for reduction of morbidity and prevention of progression disease in patients with NYHA II class having (LVEF)  $\leq$  35% and QRS  $\geq$  120 msec [3], [4].

Cardioverter-defibrillator (CD) devices were implanted for sudden cardiac prevention in patients with LVEF  $\leq$  35% after myocardial infarction and NYHA class II, III; non-ischaemic cardiomyopathy (NICMP) with LVEF  $\leq$  35% and NYHA class II and III, in patients with LVEF 30-40% after

myocardial infarction and positive electrophysiological study and for secondary prophylaxis in patients already experienced an aborted cardiac death or severe sustained ventricular tachycardia [10].

We selected for detailed presentation a case-series of 6 patients with severe ES which required hospitalization.

All the data were analysed with SPSS software (version 13.0, SPSS Inc., Chicago. IL, USA).

#### 4. Results

Most of the patients were men (190 p, 87.55%). The mean age of patients was 62.67+ 11.12 years (24-83). Seventy one patients (32.71%) were hypertensive, 132 (60.82%) had dyslipidemia, 43 (19.81%) were diabetics and 40 patients (32.71%) were smokers.

Table 1

*Summary table with patients hospitalized for electrical storm. CRT-D= cardiac resynchronization therapy with defibrillator, SC-CD= single chamber cardioverter-defibrillator, DC-CD dual chamber cardioverter-defibrillator*

No	Gender	Age	Device	Etiology for implant	Primary Prophylaxis	Time to ES (days)	No. events	Etiology for ES	EF %
1	M	65	CRT-D	Non-Ischemic cardiomyopathy	yes	740	22	hypokalemia	25
2	M	75	SC-CD	Ischemic cardiomyopathy	yes	549	>3	Beta-mimetics, aminophylline	15
3	M	79	CRT-D	Non-ischemic cardiomyopathy	yes	307	60	Pulmonary thrombo-embolism	25
4	M	63	DC-CD	Non-ischemic cardiomyopathy	no	490	17	Pocket infection, Endocarditis?	50
5	F	73	CRT-D	Ischemic cardiomyopathy	no	620	190	pneumonia	20
6	M	51	SC-CD	Ischemic cardiomyopathy	no	1	3	Ischemia?	35

The defibrillator was implanted for primary prevention of sudden cardiac death in 170 patients (78.34%) and for secondary prevention in 47 patients (21.65%). According the type of the

device, 123 pts (56.68%) received a single chamber defibrillator, 20 pts (9.21%) dual chamber defibrillator and 73 pts (33.64%) have received resynchronization therapy with defibrillation support.

All patients were treated according to current ESC guidelines [3]. Most of the patients 213 (98.2%) received a beta-blocker, 167 patients (76.9%) received a conversion enzyme inhibitor or angiotensin receptor inhibitor and spironolactone was administered in 169 patients (77.88%). Only 14 patients (6.4%) were treated with digoxin and 178 patients (82%) received a loop diuretic.

Atrial fibrillation was present in 77 patients (35%) and 19 patients (8.7%) have experienced at least one syncope before the device implantation

ES requiring hospitalization was present in 6 pts (2.76%). All patients survived the severe events.

Since every case is particular we have chosen to present each patient of these series.

**CASE 1** – AF male, 65 years, known with non-ischaemic cardiomyopathy with very dilated LV (83/70 mm) severe LV dysfunction (LVEF 22%), LBBB and ambulatory NYHA IV heart failure. CRT-D device (In Sync Sentry7298) was implanted. This is a special implantable medical device that has a complementary function apart from CRT-D ones and uses thoracic impedance to determine how much fluid is present in the thorax, such as for detecting or predicting congestive heart failure.

Two years later, after an episode of heart failure decompensation self-treated with high doses of loop diuretics (Furosemide 240 mg/day one month) he developed severe hypokalaemia of 2.5 mmol/l followed by ES consisted in many arrhythmic events, some of them followed by syncope and/or shock. Device interrogation revealed the presence of 22 arrhythmic events (10 VF and 12 FVT for which more than one therapy was attempted). The patient received in total 19 shocks in 24 hours for VT and had multiple syncope. The charge time was > 16 seconds for last events, which consist an indication for device replacement. After the normalization of potassium level and heart

failure decompensation treatment, a new device (Lumax 300 HF-T) was implanted with no other significant arrhythmic events since last FUP.

**CASE 2** – NC, 75 years, with diabetes mellitus and dyslipidaemia, previous myocardial infarction and surgical revascularization. The patient was implanted with a single chamber ICD for ischaemic cardiomyopathy with LV dysfunction (LVEF of 35%). The CRT criteria were not accomplished. Twenty eight days after the implantation procedure, the patient was re-admitted presenting severe dyspnoea and pulmonary oedema. The transthoracic echocardiography revealed a severe LV dysfunction with LVEF of 15%. On ECG multiple episodes of VT with 150 beats/min. Device interrogation showed multiple episodes of VT with 150-160 beats/min. The device has not performed any intervention because the frequency of VT was below the detection limit. The therapy limits were 171 bpm for VT1, 200 bpm for VF and 231 bpm for fast VF.

The dyspnea was interpreted as bronchus spasm and the patient was treated with inhalator  $\beta$ -agonists and aminophylline which probably triggered the ES.

Under intensive treatment of heart failure, the outcome was good, with no arrhythmic events thereafter.

**CASE 3** – VZ, 79 years, no cardiovascular risk factors, was implanted with a CRT-D for non-ischemic dilated cardiomyopathy. At the moment of implantation, the LVEF was 25% with a very dilated LV (80/57 mm). Eight months after the implantation, he has a surgery procedure for gallstones. Nine days after he was admitted in our clinic after he received multiple shocks. At clinical examination we found severe dyspnea and left leg deep thrombosis suggesting. Transthoracic echocardiography showed mild pulmonary hypertension and severe

LV dysfunction (LVEF of 25%). A thoracic CT scan confirmed pulmonary thromboembolism. The outcome was favourable under complex therapy, including anticoagulant and amiodarone, with no arrhythmic events thereafter.

**CASE 4** - GA, 63 years with cardiovascular risk factors (hypertension, dyslipidemia, cigarette smoking). The underline pathology was coronary artery disease treated with PTCA with stent implantation on ACX. He had no LV dysfunction, but he experienced multiple episodes of VT/VF after revascularization therapy. The patient was implanted with a dual chamber CD (Lumax 300 DR-T) for secondary prophylaxis of SCD.

Eleven months after the device implantation he came to ambulatory clinic because he received a shock. Device interrogation showed fracture of the defibrillator lead. The lead was eventually replaced and the defibrillation threshold was tested. The procedure was complicated with pocket hematoma. The pocket content was evacuated, but after three days the patient started to present chills and fever. The pocket inspection showed deep infection, thus the device was explanted. Amethicillin resistant staphylococci was isolated from the pocket and the infection was treated with antibiotics after germ sensibility was tested. After 7 days a sterile system was re-implanted at the same part.

One month later the patient came again with ES consisted in 17 arrhythmic events (VT, VF) treated with ATP and 2 shocks. Blood cultures were positive for methicillin resistant staphylococci. The antibiotic treatment was started. After one week of surveillance, with no new arrhythmic episodes, the patient was transferred in an infectious disease department. After a very long course of antibiotic treatment, the whole system was explanted in a dedicated centre and, another device was then implanted via right subclavian vein.

**CASE 5** - RG female, 73 years. In February 2013 after the two episodes of aborted sudden death due to ventricular fibrillation, she was evaluated in our department. At that time, ECG revealed sinus rhythm with 70 beats/min., right bundle branch block left posterior bundle branch block and PR prolongation of 260 msec. Echocardiography showed systolic dysfunction with a LVEF of 35 %. The coronary angiography revealed significant stenosis on mid left anterior descending artery and thus, percutaneous angioplasty with stent implantation (XIENCE PRIME LL 3,5X33mm) was performed. According to guidelines, CRT-D therapy was indicated and a St Jude Medical PROMOTE™ Plus device was implanted. We have tested the defibrillation threshold and we found values under 20 J.

In November 2013 she was again referred to our department after a long hospitalization (30 days) for severe pneumonia in an internal medicine department. During this hospitalization, she was in medical intensive care unit for 20 days. Antibiotic and general supportive treatment was given, with resolution of pneumonia, but deterioration of her cardiovascular condition. During her stay in internal medicine unit, continuous cardiac monitoring was unfortunately not possible, still serial ECG recordings were performed as needed.

At admission in our clinic, the patient was in a very severe condition, with heart failure decompensation, class NYHA IV.

Echocardiography showed mitral regurgitation grade III, severe systolic LV dysfunction with LVEF of 20% and pulmonary hypertension of 60 mm Hg.

The device interrogation showed EOL of the device VVI resetting and the event history was not available. The device was eventually sent to headquarter producer St Jude Medical. The device was interrogated and has found to be in Backup VVI

(BVVI) mode. Analysis of the image revealed the device charged 190 times in approximately 90 minutes prior to the device reset to BVVI mode.

After few more days of treatment, we decided to replace the generator with a new one and thus, a new PROMOTE™ Plus device was implanted uneventfully. The outcome was favourable with no more arrhythmic events.

**CASE 6** – FA, male, 52 years, ischaemic cardiomyopathy, old myocardial infarction with no possible revascularization therapy. He was evaluated after one symptomatic sustained VT episode cardioverted with DC shock. Echocardiography showed LV dysfunction with LVEF of 35% and no criteria for resynchronization therapy. A single chamber CD was implanted. Next day he presented 3 episodes of VT treated with shock (1 episode) and ATP the other two episodes. We have added amiodarone on top of anti-ischaemic therapy. The patient was discharged with no further events and remained without arrhythmia after one month follow-up.

## 5. Discussions

Electrical storm is an important, risk factor for subsequent death among ICD recipients, particularly in the first 3 months after its occurrence [5].

Predictors of ES are: chronic heart failure, secondary prophylaxis of SCD (device implantation for VT, VF), low LVEF (<25%), wide QRS (>120 msec), use of digoxin, absence of beta-blocker therapy, absence of revascularization therapy after index arrhythmia, coronary artery disease [1], [6], [8].

Apart from these factors, the common triggers for ES are ischemia, electrolyte disturbances, heart failure and some others that remained unidentified.

The immediate consequence of an ES is hospitalization, particularly in patients who have received a shock from the device. Apart from prognostic implications, electric instability induces a status of anxiety with

psychological repercussion and may severely affect patient's quality of life [2].

A patient with ES has to be hospitalized in an intensive care unit. The most urgent evaluation concerns the hemodynamic stability of the arrhythmias and, if they degenerate into acute heart failure, prompt assessment of the complications linked to this (such as pulmonary edema or acute renal insufficiency). Whenever a trigger is identified, we have to take prompt measures to correct it.

In case-series presented in this paper, in one case (1) the trigger was hypokalemia and after the correction of potassium level, the outcome was good.

In two cases we have had an infectious trigger (a pocket infection - case 4 and a severe pneumonia – case 5). In the first case, the infection was impossible to be overcome, so the whole system had to be removed. In the second case, the severe electrical storm led to device exhaustion. After a complex treatment and new device implanted, the outcome was uneventfully for next six months.

An ischemic trigger was probable present in case 6, unfortunately with no revascularization solution. In this case higher dose beta-blocker and amiodarone were sufficient to limit the ES.

In case 3, general surgery was complicated with pulmonary thromboembolism which probably led to aggravation of heart failure and ES.

In case 1, 3 and 6 amiodarone was added on top of standard therapy. In the absence of contraindications (such as QT lengthening or polymorphic ventricular tachycardia), amiodarone is generally the antiarrhythmic drug of choice and has been validated in many clinical trials [7], [9].

## 6. Conclusions

Early detection and appropriate treatment of every ES episode is very important since ES represents an important, independent marker for subsequent death among ICD recipients, particularly in the first 3 months after its occurrence [6].

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