Review Article

Electrical Storm: A New Challenge in the Age of Implantable Defibrillators

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Automatic implantable cardioverter/defibrillators (AICD) increase the life expectancy of patients with underlying cardiac disease in both the primary[1,2] and secondary[3,4] prevention of sudden cardiac death (SCD). Unfortunately, despite the continuing technological development of these devices, a large number of patients experience unpleasant effects caused by their defibrillator.[4] Frequent discharges in response to incessant or recurrent ventricular tachycardia or fibrillation, the delivery of inappropriate shocks in the absence of ventricular tachycardia because of non life-threatening supraventricular tachycardias or environmental factors, are the most common negative events that can occur after implantation of the defibrillator.[4]

Definition of electrical storm

Electrical storm has been defined[5-8] as the occurrence of ventricular tachycardia or fibrillation at frequent intervals, namely three or more episodes within 24 hours, and it is definitely an emergency medical condition.[9-12] An alternative definition of electrical storm proposed recently is the occurrence of at least two episodes of ventricular tachyarrhythmia within 24 hours.[13,14] Although this arrhythmiological complication has been known for years and was associated with increased mortality before the AICD era –especially during the acute phase of myocardial infarction, in drug toxicity, or after cardiac surgery– it is particularly relevant today because of the enhanced life expectancy of the increasing number of high-risk cardiac patients who are treated with an AICD.[10] Fifty to seventy percent of patients who are given a defibrillator for the secondary prevention of SCD receive appropriate therapy at two years,[9] and at three years 10-20% will have experienced electrical storm.[5-7,12,14] Multiple discharges create psychological problems for patients and have a negative impact on their quality of life.[11] The continually increasing indications for defibrillator implantation[1-4] make electrical storm an interesting issue to resolve, not only because of its emergency nature and the psychological problems associated with it, but also because of its unfavourable effect on the patient's long-term prognosis.[7,12]

Clinical and laboratory characteristics of patients with electrical storm

What are the characteristics that predispose a patient to the occurrence of this phenomenon? From a recent study it appears that patients with severely compromised left ventricular function, chronic renal failure, and ventricular tachycardia as

initial arrhythmia have the greatest probability of experiencing electrical storm.[13] The majority (90%) of patients in that study had ventricular tachycardia, while ventricular fibrillation occurred in 8%. A small percentage had *torsades de pointes* and polymorphic ventricular tachycardia. It is noteworthy that, in spite of a detailed analysis of the electrocardiogram, haematological and biochemical examinations, and the patients' clinical symptoms, in only 36% was any triggering mechanism found that could provoke electrical storm. Those factors were ischaemia as acute coronary syndrome, infection with high fever, hypokalaemia or hyperkalaemia, hyperthyroidism, and acute heart failure. This interesting clinical observation was confirmed by all retrospective and prospective studies carried out of patients with an AICD who experienced electrical storm.[5,7,12,14,15] Thus, this study concluded that the patients with a defibrillator who are likely to undergo electrical storm are those who have a low left ventricular ejection fraction (LVEF) and chronic renal failure.[13] The same conclusions were also reached by older studies.[7] It is possible that the coexistence of advanced stage heart failure, as well as older age, could be more powerful prognostic indexes than LVEF.[12]

In contrast, patients with ventricular fibrillation as first arrhythmia and those who are taking hypolipidaemic medication have a smaller probability of developing electrical storm.[13] The AVID study[16] confirmed that there are fewer episodes of ventricular tachycardia in defibrillator patients who presented with ventricular fibrillation than in those who initially had ventricular tachycardia. However, in a recent, retrospective study with a large patient population a significant percentage of patients with electrical storm had ventricular fibrillation as first presenting arrhythmia in their history.[14] That study, unlike other observational studies, found that in a significant percentage of cases of electrical storm the arrhythmia that was detected and terminated by the AICD was not ventricular tachycardia but ventricular fibrillation. One possible reason for this is the very low LVEF. The lower the LVEF, the greater the likelihood of ventricular fibrillation and SCD.

It has been suggested that diabetes mellitus exerts a paradoxical protective effect[13] against electrical storm. However, in the recent SCD-HeFT study[17] an examination of its correlation with prophylactic defibrillator implantation in patients with LVEF <35% revealed that non-diabetic patients had a greater benefit than diabetics, although they did not show a lower risk of electrical storm. Thus, this observation needs to be supported by future studies before it can be accepted.

Electrical storm has been described in patients with post-infarction coronary artery disease, as well as in patients with various forms of cardiomyopathy, valvular disease, surgically corrected congenital heart disease, and genetically determined cardiac diseases without any apparent underlying structural disease, such as Brugada syndrome.[12] In one study the underlying coronary artery disease was an independent risk factor for electrical storm.[14] It appears that this life-threatening arrhythmiological complication follows the clinical and laboratory profile of the high-risk cardiac patient who is treated with an AICD for protection against SCD. Although in the series of patients with electrical storm published so far in the literature this complication has mainly been described in patients with a previous history of terminated sustained ventricular tachyarrhythmia (secondary prevention of SCD),[5-7,12,13,15,18] it is also likely to occur in high-risk patients who are treated with an AICD for primary prevention of SCD.[14,19] In those patients it has been found that the combination of a very low LVEF (≤25%) with a wide QRS complex (≥120 ms) is a powerful prognostic factor for the occurrence of electrical storm.[19] For this reason these patients should undergo frequent clinical and laboratory monitoring in order to reduce the probability of occurrence or reoccurrence of electrical storm.

Long-term prognosis of patients with electrical storm

Electrical storm is probably a bad prognostic factor for long-term outcome,[7,12,14,18] although some investigators maintain that it is not a marker of increased future mortality.[5,6,13] These disagreements are due to the different patient populations studied, different definitions of electrical storm, the retrospective or prospective collection of data, and the different follow-up times (Table 1). Thus, in the studies that did not find an unfavourable long-term outcome either the follow-up duration was shorter or the definition of electrical storm was looser.[5,13] The consistent finding from all observational studies in which electrical storm was an unfavourable prognostic factor was that the increased mortality was due to rapidly deteriorating heart failure, with a small proportion of sudden cardiac or other deaths.[7,12,15,18] Indeed, in one prospective observational study of patients treated with an AICD for malignant ventricular arrhythmias, the independent risk factors for increased mortality from

Table 1. Prognostic significance of electrical storm in patients with an implantable defibrillator. The "Definition" column shows the number of episodes of ventricular tachycardia or fibrillation that were deemed by the investigators to constitute electrical storm.

Author (ref.)	Patients	Electrical storm (%)	Definition (VT/VF)	Follow up (months)	Long-term outcome
Villacastin[18]	80	16 (20%)	3/24 hr	21 ± 19	Negative
Credner[5]	136	14 (10%)	3/24 hr	13 ± 7	Neutral
Greene[6]	222	40 (18%)	3/24 hr	34 ± 31	Neutral
Exner[7]	457	90 (19%)	3/24 hr	31 ± 13	Negative
Verma[14]	2028	208 (10%)	2/24 hr	22 ± 5	Negative
Gatzoulis[12]	169	32 (19%)	3/24 hr	33 ± 26	Negative
Brigadeau[13]	307	123 (40%)	2/24 hr	28 ± 10	Neutral

pump failure during a three-year follow up were firstly an advanced stage of heart failure, followed by the occurrence of electrical storm.[12] Such an observation has important clinical and therapeutic implications for the future treatment of these patients. In another short (one-year) prospective study of patients with at least one cardioverted episode of ventricular arrhythmia, electrical storm occurred in 23% of cases and was the cause of frequent hospitalisations.[15]

In most cases these events are due mainly to episodes of ventricular tachycardia that are treated with antitachycardiac pacing and electrical discharges (Figure 1). Usually we do not find a specific cause. Given that antitachycardiac pacing is able to terminate life-threatening episodes of sustained ventricular tachycardia silently, as soon as they start, without delivering defibrillatory shocks, the true incidence of electrical storm has probably been underestimated.[15] Thus it is likely that these patients, like those who experience a single electrical discharge as well as other, separate episodes of subclinical termination of ventricular tachycardia by antitachycardiac pacing, may not seek medical assistance or hospital treatment. It remains unknown to what extent the number of episodes of sustained ventricular tachyarrhythmia terminated by the AICD during electrical storm contains prognostic information regarding the patient's future outcome. It seems, however, that the type of ventricular tachyarrhythmia involved (tachycardia or fibrillation) during electrical storm does not affect the long-term outcome in these patients.[14]

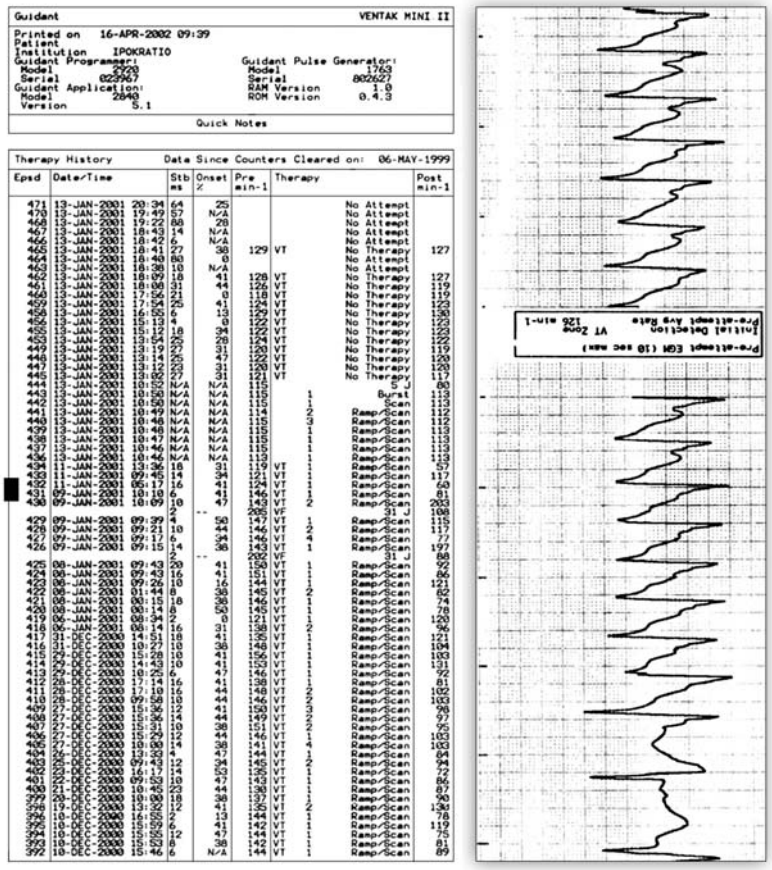
Treatment of patients with electrical storm

The patient who exhibits electrical storm should be examined very carefully and should undergo thorough clinical and laboratory testing in an attempt to determine the cause that triggered the arrhythmia, such as electrolyte disturbances or recurrent ischaemia. If one is found, we treat the underlying cause, often without the need for a specific antiarrhythmic drug.[20] Unfortunately, however, as mentioned above, such a correctible factor is only found in a few cases.

As far as antiarrhythmic medication is concerned, the choice depends on the underlying cardiac disease, on the existence and severity of heart failure, and on the chance of side effects from the drug in each individual patient.

Amiodarone remains the first choice drug in these patients because of its high efficacy and few side effects when administered over a short time. After resuscitation it is preferable to administer it intravenously.[21] Sotalol is another attractive choice that has a better side-effect profile in long-term follow up.

Class I antiarrhythmic drugs are used widely, with variable success rates, and can play a role in polymorphic ventricular arrhythmias.[22] Beta-blockers have shown good effects on both heart failure and the sympathetic system.[19] Light sedation or general anaesthesia may be needed in exceptionally resistant cases for the immediate treatment of electrical storm.[23] It is likely that the combination of amiodarone with a beta-blocker, and in particularly resistant cases with the addition of mexiletine, could be an effective therapeutic choice.[12] Indeed, in a one-year prospective follow up of high risk cardiac patients with a history of sustained ventricular tachyarrhythmia and an AICD (the Optimal Pharmacological Therapy In Cardioverter-defibrillator patients –OPTIC– trial) electrical discharges were delivered in 38.5% of patients who were taking beta-blockers, in 24.3% of those on sotalol, and in only 10.3% of patients who were taking amiodarone in combination with a beta-blocker.[24] The optimum combination of beta-blockers and angiotensin inhibitors, mainly in high-risk patients (LVEF <25% and QRS ≥120 ms),



Guidant VENTAK MINI II

Printed on 16-APR-2002 09:39
Patient
Institution IPOKRATIO
Guidant Programmer:
Model 2720
Serial 022967
Guidant Application:
Model 2048
Version 5.1

Guidant Pulse Generator:
Model 1763
Serial 802627
RAM Version 1.0
ROM Version 0.4.3

Quick Notes

Therapy History — Data Since Counters Cleared on: 06-MAY-1999

Epsd	Date/Time	Stb ms	Onset %	Pre min-1	Therapy		Post min-1
471	13-JAN-2001 20:34	64	25			No Attempt	
470	13-JAN-2001 19:49	57	N/A			No Attempt	
468	13-JAN-2001 19:22	68	28			No Attempt	
467	13-JAN-2001 18:43	14	N/A			No Attempt	
466	13-JAN-2001 18:42	6	N/A			No Attempt	
465	13-JAN-2001 18:41	27	38	129	VT	No Therapy	127
464	13-JAN-2001 18:40	80	0			No Attempt	
463	13-JAN-2001 18:38	10	N/A			No Attempt	
462	13-JAN-2001 18:09	18	41	128	VT	No Therapy	127
461	13-JAN-2001 18:08	31	44	126	VT	No Therapy	119
460	13-JAN-2001 17:56	21	0	118	VT	No Therapy	119
459	13-JAN-2001 17:54	25	41	124	VT	No Therapy	123
458	13-JAN-2001 16:55	6	13	122	VT	No Therapy	130
456	13-JAN-2001 15:13	4	0	122	VT	No Therapy	123
455	13-JAN-2001 15:12	18	34	122	VT	No Therapy	123
453	13-JAN-2001 13:54	25	28	124	VT	No Therapy	122
449	13-JAN-2001 13:19	27	31	120	VT	No Therapy	119
448	13-JAN-2001 13:14	25	47	122	VT	No Therapy	120
447	13-JAN-2001 13:12	23	31	120	VT	No Therapy	120
445	13-JAN-2001 13:02	27	31	121	VT	No Therapy	117
444	13-JAN-2001 10:52	N/A	N/A	115		5 J	80
443	13-JAN-2001 10:50	N/A	N/A	115	1	Burst	113
442	13-JAN-2001 10:50	N/A	N/A	115	1	Scan	113
441	13-JAN-2001 10:49	N/A	N/A	114	2	Ramp/Scan	112
440	13-JAN-2001 10:48	N/A	N/A	115	3	Ramp/Scan	112
439	13-JAN-2001 10:48	N/A	N/A	115	1	Ramp/Scan	113
438	13-JAN-2001 10:47	N/A	N/A	115	1	Ramp/Scan	113
437	13-JAN-2001 10:46	N/A	N/A	115	1	Ramp/Scan	113
436	13-JAN-2001 10:46	N/A	N/A	113	1	Ramp/Scan	113
434	11-JAN-2001 07:36	18	31	119	VT 1	Ramp/Scan	57
433	11-JAN-2001 07:15	14	34	121	VT 1	Ramp/Scan	117
432	11-JAN-2001 06:17	16	41	124	VT 1	Ramp/Scan	60
431	09-JAN-2001 09:10	6	41	144	VT 1	Ramp/Scan	81
430	09-JAN-2001 10:09	10	47	143	VT 2	Ramp/Scan	203
		2	--	205	VF 1	31 J	108
429	09-JAN-2001 09:39	4	58	147	VT 1	Ramp/Scan	115
428	09-JAN-2001 09:21	10	44	146	VT 2	Ramp/Scan	117
427	09-JAN-2001 09:17	6	34	146	VT 4	Ramp/Scan	77
426	09-JAN-2001 09:15	14	38	143	VT 1	Ramp/Scan	197
		2	--	202	VF	31 J	88
425	08-JAN-2001 09:43	20	41	150	VT 1	Ramp/Scan	92
424	08-JAN-2001 09:43	16	41	151	VT 1	Ramp/Scan	86
423	08-JAN-2001 09:26	10	16	144	VT 1	Ramp/Scan	121
422	08-JAN-2001 01:44	8	38	145	VT 2	Ramp/Scan	82
421	08-JAN-2001 00:15	10	38	146	VT 1	Ramp/Scan	74
420	08-JAN-2001 00:14	8	50	145	VT 1	Ramp/Scan	78
419	06-JAN-2001 08:34	2	0	121	VT 1	Ramp/Scan	120
418	06-JAN-2001 08:14	16	31	138	VT 2	Ramp/Scan	96
417	31-DEC-2000 14:51	18	41	135	VT 1	Ramp/Scan	121
416	31-DEC-2000 09:27	10	38	148	VT 1	Ramp/Scan	104
415	29-DEC-2000 15:28	10	41	156	VT 1	Ramp/Scan	103
414	29-DEC-2000 14:43	10	41	153	VT 1	Ramp/Scan	131
413	29-DEC-2000 10:25	6	47	146	VT 1	Ramp/Scan	92
412	28-DEC-2000 17:14	16	41	138	VT 1	Ramp/Scan	81
411	28-DEC-2000 17:10	16	44	148	VT 2	Ramp/Scan	102
410	28-DEC-2000 09:58	10	44	146	VT 2	Ramp/Scan	103
409	27-DEC-2000 15:36	12	41	150	VT 3	Ramp/Scan	98
408	27-DEC-2000 15:36	14	44	149	VT 2	Ramp/Scan	97
407	27-DEC-2000 15:31	10	38	151	VT 2	Ramp/Scan	95
406	27-DEC-2000 15:29	12	44	146	VT 1	Ramp/Scan	103
405	27-DEC-2000 15:08	14	38	141	VT 4	Ramp/Scan	103
404	26-DEC-2000 13:23	4	47	144	VT 1	Ramp/Scan	84
403	25-DEC-2000 09:43	12	34	145	VT 2	Ramp/Scan	94
402	24-DEC-2000 11:17	14	53	135	VT 1	Ramp/Scan	72
401	22-DEC-2000 09:53	10	47	143	VT 1	Ramp/Scan	86
400	21-DEC-2000 10:48	23	44	138	VT 1	Ramp/Scan	87
399	20-DEC-2000 10:00	18	38	137	VT 1	Ramp/Scan	90
398	19-DEC-2000 13:32	12	41	135	VT 2	Ramp/Scan	130
396	18-DEC-2000 16:55	2	13	144	VT 1	Ramp/Scan	78
395	18-DEC-2000 15:57	6	41	142	VT 1	Ramp/Scan	112
394	18-DEC-2000 15:55	12	47	144	VT 1	Ramp/Scan	75
393	18-DEC-2000 15:53	8	38	142	VT 1	Ramp/Scan	81
392	18-DEC-2000 15:46	6	N/A	144	VT 1	Ramp/Scan	89

Figure 1. Patient aged 65 years, with post-infarction coronary artery disease, a history of sustained ventricular tachycardia, and an implantable defibrillator. Four years after implantation he suffered multiple episodes of sustained ventricular tachycardia. Interrogation of the generator showed that multiple episodes of sustained ventricular tachycardia had been terminated repeatedly by antitachycardiac pacing or defibrillator discharge within a period of a few days. The electrical storm was treated successfully with a combination of triple antiarrhythmic medication (amiodarone, metoprolol, mexiletine). From Gatzoulis KA: Ventricular arrhythmias: from the electrophysiological laboratory to clinical practice, in Stefanadis C (ed.): Kardiologika Themata, 2004; pp 135-156.

may reduce the chance of occurrence of electrical storm.[19]

The recently published SHIELD trial showed that azimilide is effective and helps to reduce the number of discharges, though not mortality.[15,25] Azimilide is an experimental class III antiarrhythmic drug that blocks calcium channels and prolongs the energy potential and refractory periods. A secondary analysis of the SHIELD results found that during a prospective one-year follow up azimilide significantly reduced the incidence of electrical storm in comparison with placebo.[15] That means that we shall have an extra alternative for the treatment of electrical storm if the drug becomes commercially available. In any case, from the data available to date we appear to be lagging behind in the effective treatment of these difficult conditions and perhaps new drugs in the future will give us greater and better powers for the prevention and treatment of electrical storm.[10]

Radiofrequency ablation of monomorphic ventricular tachycardia, or even of polymorphic ventricular tachycardia, is the next solution after the failure of medication.[26,27] Nowadays, it is feasible to suppress episodes of sustained ventricular tachycardia in patients with organic heart disease by endocardial ablation of the arrhythmogenic focus in one of the two ventricles in the electrophysiological laboratory.[26,27] Conventional mapping techniques, or more modern electroanatomical mapping methods, can be used with safety and likely efficacy.[28,29] Even though these methods do not usually lead to the disappearance of the arrhythmiological substrate, they may modify it sufficiently to make it difficult or impossible for a previously easily-triggered sustained ventricular tachycardia to reoccur, allowing the possibility of preventing relapses without the need for long-term combined antiarrhythmic medication (Figure 2). To what extent this approach might contribute to an improvement in the patient's life expectancy is unknown.

Every attempt should be made to achieve optimum programming of the AICD so that life-threatening episodes of ventricular tachyarrhythmias will be terminated promptly.[30,31] Finally, in certain patients, upgrading to biventricular pacing or even heart trans-

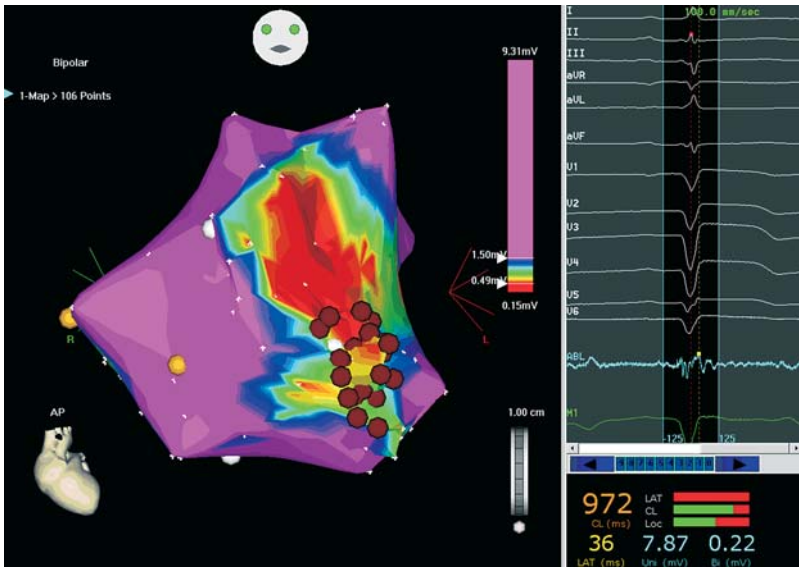


Figure 2. Patient aged 65 years, with coronary artery disease, who presented with repeated episodes of sustained ventricular tachycardia (electrical storm) prior to AICD implantation. The episodes were treated with triple antiarrhythmic medication and subsequent ablation-modification of the arrhythmogenic substrate using a three-dimensional colour electroanatomical mapping system (potential map). Three years later the patient remained in stage II heart failure and the AICD had been successfully activated with antitachycardiac pacing only once, three months after ablation and defibrillator implantation.

plantation is recommended, if there is worsening of the stage of heart failure and the necessary prerequisites are satisfied.[6,32,34]

Summation

Protection against electrical storm is difficult, but an improvement in the life expectancy of patients with advanced heart disease and an AICD is achievable today. It requires an understanding of the cause of heart failure, and a good knowledge of the antiarrhythmic and other drugs that are used for the optimum treatment of these patients. Even if all this fails, radiofrequency ablation provides an alternative solution. Ischaemia, electrolyte disturbances, proarrhythmia, drug intolerance, the use of inotropic or other agents that can trigger ventricular tachycardia or fibrillation, should all be corrected as soon as they are detected. Electrical storm is a tragic experience for the patient, with many psychological and financial consequences. Every possible attempt should be made to reduce as far as possible the number of patients who undergo it. It cannot be eliminated, but it can be mitigated.

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