

# Current Management of Electrical Storm

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The management of ventricular electrical storm can prove to be quite a daunting challenge for the clinician given its complexity and life threatening consequences. Despite the gravity of its nature, there is a surprising paucity of information currently available. Owing to a lack of consensus on its management, the clinician is often left without adequate direction and information to properly guide therapy. We review the literature and discuss potential etiologies as well as current therapeutic modalities. An algorithm is presented that offers the clinicians a guideline of how to manage the electrical storm.

**Key Words:** Arrhythmia • Electrical storm • Ventricular fibrillation • Ventricular tachycardia

## INTRODUCTION

The management of ventricular electrical storm (often simplified as electrical storm) can prove to be a challenging task for the clinician considering its critical and unstable nature. Further adding to the quandary is the lack of uniformity of its management. This review will address current understanding of the mechanisms as well as the leading treatment options for ventricular electrical storm. Included will be a detailed review of the literature, as well as a proposed algorithm on how to manage electrical storm.

### Definition and etiology of electrical storm

The most widely accepted definition for electrical storm is a recurrent condition of hemodynamically significant ventricular tachyarrhythmia occurring at least two or more times over a 24 hour time frame.<sup>1</sup> The

tachyarrhythmias are usually manifested in the form of ventricular tachycardia or ventricular fibrillation, and often require multiple electrical cardioversions. Determining the etiology of the condition, as well as its subsequent management can be quite challenging, and thus we will attempt to elucidate the etiology of electrical storm, as well as review the literature on the management strategies.

The etiology of ventricular electrical storm is fairly broad, and encompasses several different areas. Potential causes include enhanced sympathetic tone, ischemia, electrolytes, endocrine disorders, genetic abnormalities (such as Brugada syndrome, long QT syndrome, etc.), and iatrogenic causes (Table 1).

The mechanisms of electrical storm are quite complex and not well understood. Each case of electrical storm may represent different underlying cause and electrophysiologic mechanism. It has been postulated that cellular and molecular alterations can increase intracellular calcium overload and changes of the action potential duration and morphology that lead to the onset of electrical storm.<sup>2-5</sup> It is beyond the scope and focus of this review to thoroughly discuss the complicated and diverse mechanisms of electrical storm. However, the importance of the role of enhanced sympathetic tone has been well documented. There are a multitude of conditions that could lead to increased adrenergic output, including ischemia, surgery,<sup>6</sup> hyperthermia,<sup>7</sup> and even jet

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**Table 1.** Causes of electrical storm\*

1. Enhanced sympathetic tone
2. Ischemia
3. Electrolyte imbalances (potassium, magnesium)
4. Genetic abnormalities (such as Brugada syndrome, LQTS, CPVT, ERS, etc.)
5. Iatrogenic (often in the presence of implantable cardioverter-defibrillator)
6. Endocrine disorders (thyroid disorders, pheochromocytoma)

\*The causes of electrical storm (1-6) are listed in an approximately descending order of occurrence. CPVT, catecholaminergic polymorphic ventricular tachycardia; ERS, early repolarization syndrome; LQTS, long QT syndrome.

lag.<sup>8</sup> In the pathogenesis of electrical storm, the exact relationship between sympathetic tone and ischemia has not exactly been elucidated. In other words, does ischemia itself act as a trigger for electrical storm or does it potentiate a lethal arrhythmia by enhancing sympathetic tone? This raises the question of if the relationship between ischemia and increased sympathetic tone represents one of cause and effect, or rather, do they represent separate entities with different and distinct pathologies? Some authors believe that ischemia acts as a trigger for increased adrenergic output, consequently promoting the development of unstable tachyarrhythmias.<sup>9</sup> Regardless of its relationship with sympathetic tone, ischemia is an important potentially reversible cause to consider with ventricular electrical storm. As an illustration of this point, Perzanowski describes a patient who underwent coronary artery bypass surgery (CABG), and subsequently experienced over 30 shocks postoperatively. Repeat coronary angiography revealed a kinked left internal mammary artery (LIMA) graft as the likely culprit source of ischemia. A repeat surgical revision of his anatomy relieved his ventricular tachyarrhythmias.<sup>10</sup>

In addition to adrenergic tone and ischemia, endocrine disorders such as thyrotoxicosis<sup>11</sup> and pheochromocytoma<sup>12</sup> have been shown to potentiate electrical activity in the heart causing tachyarrhythmias. In a case of amiodarone induced thyrotoxicosis, Marketou explains the case of an 18 year old woman who developed electrical storm one year out from treatment with amiodarone for sustained episodes of ventricular tachycardia secondary to idiopathic dilated cardiomyopathy.<sup>11</sup> The patient's condition was refractory to treatment with large doses of propylthiouracil and propranolol. She eventually re-

quired a total thyroidectomy, which resolved her tachyarrhythmia. The patient was subsequently started on thyroid replacement and amiodarone treatment was resumed without recurrence of tachyarrhythmias.

Genetic causes of electrical storm such as Brugada syndrome, Early Repolarization Syndrome (ERS), catecholaminergic polymorphic ventricular tachycardia, long and short QT syndromes are also important to appreciate. Brugada syndrome and ERS are similar in terms of genetic linkage, age of onset, and potential for causing electrical storm.<sup>13,14</sup> Slowed heart rates in these patients have been shown to elicit the occurrence of ventricular tachyarrhythmias. The prevailing theory behind this clinical finding is that the increase in vagal tone that accompanies a slowed heart rate leads to an increase in transmural voltage gradient, which can permit the development of a window for premature beats that trigger ventricular tachyarrhythmias. Long QT Syndrome is another genetic disorder characterized by an abnormality in the ion channel function which in turn results in repolarization abnormalities. This consequently predisposes the patient to ventricular tachyarrhythmias. Long QT syndrome (LQTS) can occur in both a congenital form as well as an acquired form. In the acquired form, a medication can prolong the QT interval in the susceptible patient and trigger an arrhythmia.

It is also important to consider iatrogenic causes in the etiology of electrical storm. The incidence of electrical storm in patients with implantable cardiac defibrillators seems to be variable when reviewing the literature.<sup>15-18</sup> Previously, during the 1980s and 1990s, when patients were treated with thoracotomy ICD devices, there was a higher incidence of electrical storm in comparison to the non-thoracotomy ICD devices used today.<sup>15,16</sup> One study noted that the highest incidence of electrical storm is most commonly seen in patients treated with an ICD for secondary prevention, specifically monomorphic VT associated with structural heart disease, and is less common in patients treated for primary prevention.<sup>17</sup> It was thought the increased ambient ectopy that occurs with electrical therapy coupled with the structural and functional changes that take place over time can potentiate arrhythmias.

Although it is imperative to consider all of the above causes of electrical storm, we must also not forget to thoroughly review each patient's medication profile.

There are many agents on the market today with possible unknown arrhythmogenic effects. The importance of this consideration is shown in one case report that documents an association of electrical storm with the usage of Ginkgo biloba.<sup>19</sup>

### Management of electrical storm

After determining the etiology of this life threatening condition, the clinician must then shift the attention on effective management. However, in treating electrical storm, the clinician is often faced with a lack of direction and evidence based guidelines. Therefore, a brief review of the literature will be discussed here. Treatment can be broadly classified into two modalities: pharmacological and non-pharmacological (Table 2).

### Pharmacological therapy

#### Single drug therapy

In regards to pharmacological therapy, viable options include sympathetic tone inhibition and antiarrhythmics. Beta-antagonists are becoming increasingly utilized to attenuate enhanced sympathetic output. Other methods described in the literature targeted at reducing adrenergic tone include left stellate ganglion blockade<sup>20</sup> and propofol.<sup>21</sup> The rationale for left stellate ganglion blockade is inhibition of the sympathetic innervations from the ganglion to the left ventricle.<sup>20</sup> There is also said to be an alpha blockade effect caused by this procedure. Although usually very effective, in the acute setting this method of sympathetic blockade is not always practical, and it requires a high level of expertise in order to perform successfully. In the case of propofol, there is a case report in which boluses of propofol were administered and resolved a patient's ventricular electrical storm which had been previously refractory to antiarrhythmics.<sup>21</sup> The mechanism by which propofol attenuates adrenergic tone was thought to be derived from its actions on brain stem vasomotor centers, consequently leading to decreased sympathetic output. However, it is not always beneficial to induce sympathetic blockade in all patients with electrical storm. When managing electrical storm in patients with known ERS or Brugada syndrome, attenuation of sympathetic tone is not recommended.<sup>13,14</sup> Slowed heart rate in these conditions can actually promote electrical storm. In one case

report, a patient with ERS developed electrical storm, which was reversed by isoproterenol.<sup>14</sup> The effectiveness of isoproterenol in this case was thought to deal with its ability to enhance the inward Ca<sup>2+</sup> current, which eliminates the transmural voltage gradient that creates the window for premature beats that trigger ventricular tachyarrhythmias.

The other major arm of pharmacotherapy besides sympathetic blockade includes antiarrhythmics, which are often the first line agents deployed. Specific antiarrhythmics described in the literature include class IA agents such as amiodarone (also a Class III agent) and procainamide, Class IB agents such as lidocaine and Class III agents such as bretylium. A new experimental novel class III agents called nikelant hydrochloride which selectively blocks the rapid component of delayed rectifier K<sup>+</sup> current, has also proven useful.<sup>22</sup> The aforementioned novel agent, nikelant was described in a case report of a patient who experienced refractory ventricular tachycardia after CABG and Dor procedure. There has been a case report describing a patient with electrical storm refractory to many single agents, but responsive to bretylium.<sup>23</sup> In addition to the selection of the drug itself, there have been reports of the route of administration possibly affecting efficacy. A case report was published in where a patient with ventricular tachycardia storm was refractory to chronic oral amiodarone, but sensitive to intravenous amiodarone.<sup>24</sup> Although the authors remarked that the mechanism of this finding was not entirely clear, it is possible that reduced time to onset

**Table 2.** Treatment of electrical storm

Pharmacological
Sympathetic blockade
Beta antagonists
Left stellate ganglion blockade
Propofol
Antiarrhythmics
Combination therapy
Beta antagonist + class III antiarrhythmic
Beta antagonist + class III antiarrhythmic + class IB
Non-Pharmacological
Cather ablation
Overdrive pacing
Intraaortic balloon pump or extracorporeal life support
Heart transplant

of therapeutic effect with intravenous amiodarone as compared to oral amiodarone played a role. In addition, oral amiodarone is thought to have a greater variability in bioavailability, which is most likely attributable to its variable absorption.

With so many agents available, it may become difficult for the clinician to decide upon which agent to use first. By convention, many clinicians ultimately chose an antiarrhythmic such as intravenous amiodarone for first line monotherapy. However, the question arises whether this approach of choosing an antiarrhythmic first is based merely on convention and habit, or whether there is evidence in the literature to base this decision. One study compared the use of intravenous amiodarone with bretylium and showed that amiodarone was at least as effective and was more hemodynamically tolerable than bretylium, since it has a tendency to promote hypotension.<sup>25</sup>

A pivotal study examined the efficacy of sympathetic blockade versus antiarrhythmics in 49 patients with electrical storms.<sup>26</sup> The investigators ultimately found that the sympathetic blockade arm had a statistically significant lower mortality rate at one week. This finding seems reasonable when we consider the known cardio-protective benefit seen in post-Mi patients treated with beta blockade. Also, antiarrhythmics particularly class I agents, while helpful in the acute setting can cause negative inotropic effects, promote heart failure, and lead to fatal arrhythmias. Thus, based on limited reports in the literature, a standard approach for utilization of sympathetic blockade over antiarrhythmics as a first line agent can be argued. Furthermore, most cases of electrical storm appear to be attributable to either enhanced adrenergic tone or ischemia, and beta antagonists are effective in both of these conditions.

### Combined drug therapy

Given the unstable nature of the disease, electrical storm has often been refractory to monotherapy, and thus it is not uncommon to deploy combination therapy.<sup>18,27</sup> Successful combination therapy reported in the literature include a beta antagonist in conjunction with a class III antiarrhythmic. In reviewing the literature, it seems that the beta antagonist most often deployed was propranolol. Further adding to the support of the specific use of propranolol is a case report of electrical storm being re-

fractory to metoprolol and amiodarone, but sensitive once propranolol was substituted for metoprolol.<sup>27</sup> There have also been retrospective studies that have shown a benefit of combining class III agents such as amiodarone or sotalol with a class IC agent, flecainide. Class IC agents when used alone can be proarrhythmogenic, however this was not observed when combined with class III agents since they appeared to counter this effect.<sup>28</sup> There is also a successful case study of treating a patient using triple therapy with a beta antagonist, class III antiarrhythmic, and a class IB antiarrhythmic such as lidocaine or mexiletine.<sup>18</sup>

### Non-pharmacological therapy

When conventional pharmacotherapy fails, the clinician may turn to non-pharmacologic modalities. Such techniques reported include catheter ablation, pacing, and heart transplant. Multiple reports have described several instances of successful ablation in patients with drug refractory electrical storm.<sup>29-33</sup> However, electrical storm is not always amenable to ablative therapy since the electrical activity can be too rapid to map, and multiple pathologies morphologically may be at play. There has even been a report of drug refractory electrical storm responding to temporary overdrive AV sequential pacing.<sup>34</sup> There has also been one case study of a patient with severe left ventricular dysfunction who developed "giant electrical storm" with 90 episodes of pulseless VT prior to scheduled coronary artery bypass grafting, who responded to levosimendan and intra-aortic balloon counterpulsation.<sup>35</sup> One retrospective study of 11 patients details the use of extracorporeal life support for the termination of refractory ventricular tachyarrhythmias. By employing ECLS sufficient myocardial perfusion, and unloading of the heart was achieved, which allowed time to correct for proarrhythmogenic disturbances such as ischemic or electrolyte abnormalities.<sup>36</sup> Finally, if a patient with electrical storm has proven resistant to multiple therapies, consideration could be made then for cardiac transplant.

### Proposed algorithm for the approaches to electrical storm

Our case illustrates the biggest challenge in treating electrical storm – the lack of a standard approach or unified consensus. We were similarly faced with this di-

**Table 3.** Proposed algorithm (steps 1 to 4)

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1. Identify etiology and try to reverse any potentially reversible causes

Causes

- Enhanced sympathetic tone
- Ischemia
- Electrolytes (potassium, magnesium)
- Endocrine disorders (thyroid disorders, pheochromocytoma)
- Genetic abnormalities (such as Brugada syndrome, LQTS, etc.)
- Iatrogenic

Consider obtaining

- Electrolytes
- Ischemia work up
- Thyroid studies
- Careful review of medication history

2. Mono-Pharmacotherapy

First Line: B-blocker<sup>26</sup>

Consider using propranolol as beta antagonist of choice<sup>27</sup>

If patient is intubated and on sedation, considering using propofol to enhance sympathetic tone attenuation<sup>21</sup>

3. If mono-therapy is unsuccessful → Combination therapy

B-Blocker + class III Antiarrhythmic<sup>28</sup>

If amiodarone is chosen, consider using intravenous form<sup>24</sup>

B-Blocker + class III Antiarrhythmic + class IB antiarrhythmic<sup>19</sup>

4. Non-pharmacotherapy for drug refractory cases

Catheter ablation<sup>29-33</sup>

Overdrive pacing<sup>34</sup>

Intraaortic balloon pump<sup>35</sup> or extracorporeal life support<sup>36</sup>

Heart transplant

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lemma, as standard texts were limited and did not provide an adequate direction by which to base and guide our treatment. Therefore, after a careful review of the literature, the following proposed algorithm has been formulated (Table 3). It should be noted that the proposed algorithm for the treatment of electrical storm is based mainly on limited case reports and studies since there is a paucity of large randomized trials. We hope that this will provide the clinician with a useful guidance.

## REFERENCES

1. Dorian P, Cass D. An overview of the management of electrical storm. *Can J Cardiol* 1997;13:13A-7A.
2. Brooks WW, Conrad CH, Morgan JP. Reperfusion induced arrhythmias following ischaemia in intact rat heart: role of intracellular calcium. *Cardiovasc Res* 1995;29:536-42.
3. Tribulova N, Seki S, Radosinska J, et al. Myocardial Ca<sup>2+</sup> handling and cell-to-cell coupling, key factors in prevention of sudden cardiac death. *Can J Physiol Pharmacol* 2009;87:1120-9.
4. Chan YH, Wu CT, Yeh YH, Kuo CT. Reappraisal of Luo-Rudy dynamic cell model. *Acta Cardiol Sin* 2010;26:69-80.
5. Yukiomi T, Mayumi H, Niels V, et al. Ca<sup>2+</sup>-related signaling and protein phosphorylation abnormalities play central roles in a new experimental model of electrical storm. *Circulation* 2011;123:2192-203.
6. Schmidt TD, Muir AJ. A case of electrical storm in a liver transplant patient. *Transplant Proc* 2003;35:1437-8.
7. D'Aloia A, Faggiano P, Brentana L, et al. Recurrent ventricular fibrillation during a febrile illness and hyperthermia in a patient with dilated cardiomyopathy and automatic implantable cardioverter defibrillator. An example of electrical storm. *Int J Cardiol* 2005;103:207-8.
8. Han SW, Kim YN, Lee YS, et al. An electrical storm with more than 3000 shocks in patient with an ICD. Is jet-lag a trigger? *Int J Cardiol* 2005;104:235-7.
9. Kurisu S, Inoue I, Kawagoe T, et al. Temporary overdrive pacing as an adjunct to antiarrhythmic drug therapy for electrical storm in acute myocardial infarction. *Circulation* 2005;69:613-6.
10. Perzanowski C, Pai S. Electrical storm after CABG due to a kinked LIMA. *Pacing Clin Electrophysiol* 2004;27:545-6.

11. Marketou M, Simantirakis E, Manios E, Vardas P. Electrical storm due to amiodarone induced thyrotoxicosis in a young adult with dilated cardiomyopathy: thyroidectomy as the treatment of choice. *Pacing Clin Electrophysiol* 2001;24:1827-8.
12. Michaels RD, Hays JH, O'Brian JT, Shakir KM. Pheochromocytoma associated ventricular tachycardia blocked with atenolol. *J Endocrinol Invest* 1990;11:943-7.
13. Grande A, Tabita V, Lizzio MM, et al. Early repolarization syndrome and Brugada syndrome" Is there any linkage? *Eur J Intern Med* 2008;19:236-40.
14. Bernard A, Genee O, Grimard C, et al. Electrical storm reversible by isoproterenol infusion in a striking case of early repolarization. *J Interv Card Electrophysiol* 2009;25:123-7.
15. Credner S, Klingenhoben T, Mauss O, et al. Electrical storm in patients with transvenous implantable cardioverter-defibrillators. *J Am Coll Cardiol* 1998;32:1909-15.
16. Kim SG, Ling J, Fisher JD, et al. Comparison and frequency of ventricular arrhythmias after defibrillator implantation by thoracotomy versus nonthoracotomy approaches. *Am J Cardiol* 1994;74:1245-8.
17. Stuber T, Eigenmann C, Delacretaz E. Characteristics and relevance of clustering ventricular arrhythmias in defibrillator recipients. *Pacing Clin Electrophysiol* 2005;28:702-7.
18. Manolis A, Katsivas A, Vassilopoulos C, Tsatiris C. Electrical storm in an ICD-recipient with 429 delivered appropriate shocks: therapeutic management with antiarrhythmic drug combinations. *J Interv Card Electrophysiol* 2002;6:91-4.
19. Pfister O, Stichlerling C, Schaer B, Osswald S. Electrical storm caused by complementary medication with ginkgo biloba extract. *Am J Med* 2008;121;11:e3-4.
20. El-Sherif N. The challenge of management of electrical storm out-of-hospital cardiac arrest. *Cardiol J* 2007;14:326-8.
21. Burjorjee J, Milne B. Propofol for electrical storm; a case report of cardioversion and suppression of ventricular tachycardia by propofol. *Can J Anaesth* 2002;49:973-7.
22. Sahara M, Sagara K, Yamashita T, et al. Nifekalant HCL, a novel class III antiarrhythmic agent suppressed postoperative recurrent VT in a patient undergoing CABG and the Dor Approach. *Circulation* 2003;67:712-4.
23. Kapoor J, Batsford W, Foody J. Refractory electrical storm suppression by bretylium. *Int J Cardiol* 2006;113:E111-2.
24. Kavesh N, Olsovsky M, Freudenberg R, et al. IV Amiodarone suppression of electrical storm refractory to chronic oral amiodarone. *Pacing Clin Electrophysiol* 1999;22:665-7.
25. Kowey PR, Levine JH, Herre JM, et al. Randomized, double-blind comparison of intravenous amiodarone and bretylium in the treatment of patients with recurrent, hemodynamically destabilizing ventricular tachycardia or fibrillation. *Circulation* 1995; 92:3255-63.
26. Nadernanee K, Taylor R, Bailey W, et al. Treating electrical storm: sympathetic blockade versus advanced cardiac life support. *Circulation* 2000;102:742-7.
27. Tsagalou EP, Kanakakis J, Rokas S, Anastasiou-Nana MI. Suppression by propranolol and amiodarone of an electrical storm refractory to metoprolol and amiodarone. *Int J Cardiol* 2005; 99:341-2.
28. Fuchs T, Groysman R, Meilichov I. Use of a combination of class III and class Ic antiarrhythmic agents in patients with electrical storm. *Pharmacotherapy* 2008;28;1:14-9.
29. Schreieck J, Zrenner B, Deisenhofer I, Schmitt C. Rescue ablation of electrical storm in patients with ischemic cardiomyopathy: a potential-guided ablation approach by modifying substrate of intractable, unmappable ventricular tachycardias. *Heart Rhythm* 2005;2:10-4.
30. Bänsch D, Oyang F, Antz M, et al. Successful catheter ablation of electrical storm after myocardial infarction. *Circulation* 2003; 108:3011-6.
31. Kolettis TM, Naka KK, Katsouras CS. Radiofrequency catheter ablation for electrical storm in a patient with dilated cardiomyopathy. *Hellenic J Cardiol* 2005;46:366-9.
32. Carbuicchio C, Santamaria M, Trevisi N, et al. Catheter ablation for the treatment of electrical storm in patients with implantable cardioverter-defibrillators: short- and long-term outcomes in a prospective single-center study. *Circulation* 2008;117:462-9.
33. Tanner H, Hindricks G, Volkmer M, et al. Catheter ablation of recurrent scar-related ventricular tachycardia using electroanatomical mapping and irrigated ablation technology: results of the prospective multicenter Euro-VT-Study. *J Cardiovasc Electrophysiol* 2010;21:47-53.
34. Kurisu S, Inoue I, Kawagoe T, et al. Temporary overdrive pacing as an adjunct to antiarrhythmic drug therapy for electrical storm in acute myocardial infarction. *Circulation* 2005;69:613-6.
35. Vaneka T, Kolesara M, Nejedlya M, Jirmarb R. Rescue peri-operative management of the patient with giant electrical storm and severe left ventricular dysfunction: support by levosimendan and intraaortic balloon counterpulsation. *Interact Cardiovasc Thorac Surg* 2008;7:648-50.
36. Tsai FC, Wang YC, Huang YK, et al. Extracorporeal life support to terminate refractory ventricular tachycardia. *Crit Care Med* 2007;35:1673-6.