

# Management of an Electrical Storm: Definitions, Mechanisms, Diagnosis, and Treatment Strategies

Antônio da Silva Menezes Júnior<sup>1,2\*</sup>, Ana Luisa Adorno de Lima<sup>1</sup>, Roberta Gomes da Mata<sup>1</sup>, Dayanne Cardoso Teixeira<sup>1</sup>

<sup>1</sup> School of Medical, Pharmaceutical, and Biomedical Sciences, Pontifical Catholic University of Goiás, Goiânia, Brazil

<sup>2</sup> Federal University of Goiás, Faculty of Medicine, Brazil

\* **Corresponding Author:** Antônio da Silva Menezes Júnior, School of Medical, Pharmaceutical and Biomedical Sciences, Pontifical Catholic University of Goiás, Rua 235, number 1440, SetorLesteUniversitário, GoiâniaGoiás, Brazil. E-mail: [a.menezes.junior@uol.com.br](mailto:a.menezes.junior@uol.com.br)

Received April 3, 2021; Accepted May 19, 2021; Online Published March 6, 2022

## Abstract

An Electrical Storm (ES) is a Ventricular Arrhythmia (VA) consisting of the occurrence of three or more Ventricular Tachycardias (VT) per day, separated by five-minute intervals, or the presence of unceasing VT, even with the optimization of antiarrhythmic drug therapy. Data revealed that 84 out of 1274 patients with an Implantable Cardioverter-Defibrillator (ICD) developed ES during follow-up. The incidence of ES in patients with ICD can reach 20% during the first 134 days after implantation. The pathophysiological mechanism is not well understood as ES can be caused by several clinical conditions and through several different mechanisms, including VT and Ventricular Fibrillation (VF), global acute ischemia, and myocardial dysfunction. Patients with ES have a threefold greater risk of Sudden Cardiac Death (SCD). The treatment is multimodal and consists primarily of emergency sedation, ventilation, neuraxial modulation, drug therapy (beta-blockers, amiodarone, sotalol, class I anti-arrhythmic drugs), and Catheter Ablation (CA). CA is a rescue procedure performed when there is little or no response to drug therapy. This approach is performed by first mapping the local and then choosing between approaches (endocardial or epicardial) and the different CA methods, which mainly include radiofrequency ablation, irrigated radiofrequency ablation, pulsed radiofrequency ablation, alcohol ablation, and cryoablation.

**Keywords:** Electrical Storm, Ventricular Tachycardia, Ventricular Fibrillation, Implantable Cardioverter-defibrillator, Catheter

## Introduction

An ES (electrical storm) is a type of VA (ventricular arrhythmia) consisting of the occurrence of three or more VT (ventricular tachycardia) events per day, separated by five-minute intervals, or the presence of unceasing VT, even with the optimization of antiarrhythmic drug therapy. ES is common,<sup>1-4</sup> although polymorphic VT and VF (ventricular fibrillation) can also cause it.<sup>5</sup> ES might require antiarrhythmic drug therapy and an ICD (implantable cardioverter defibrillator), which is a clinical emergency.<sup>5</sup> CA (catheter ablation) is used to permanently treat ES, especially ICD interventions, and to improve patients' quality of life.<sup>6,7</sup>

## Epidemiology

A prospective randomized study was conducted, consisting of 633 patients with one documented VT or VF episode that occurred  $\leq 42$  days before the first ICD implant or who already had an ICD implant and received a shock, probably because the ICD was

triggered by VT or VF. From among the 633 patients, ES was present in 148 (23.3%) patients, with a mean age of 62 years ( $\pm 11$  years), 105 (71%) had Ischemic Heart Disease (IHD), 58 (39%) had vascular disease, 95 (64%) had previous Myocardial Infarction (MI), 102 had congestive heart failure (69%), and 76 (51%) had idiopathic dilated cardiomyopathy. ICD was indicated for VF in 30 patients (20%) and VT in 118 patients (80%).<sup>8</sup>

A Japanese study focused on 1274 patients receiving ICD therapy for structural heart disease. The underlying diseases included IHD (482 patients, 38%), dilated cardiomyopathy (342, 27%), hypertrophic cardiomyopathy (204, 16%), arrhythmogenic right ventricular cardiomyopathy (29, 2%), and other (218, 17%). ES occurred in 84 of the patients (6.6%) in this study.<sup>9</sup>

Considering this, in patients who underwent ICD implantation, the incidence of ES was high during the first two years after the procedure but varied between

10% and 20% during the first 134 days.<sup>10,11</sup> There is also a global incidence that ranges from 10%-30% at 4-9 months after the procedure when compared between larger study populations, and ES is more prevalent in males.<sup>9,11,12</sup>

In cases of primary prevention of sudden death, the incidence of ES is 4%, while for secondary prevention, it is 10%-40% in non-chagasic populations.<sup>13,14</sup>

### Physiopathology

The ES mechanism has not been fully elucidated, as it can be triggered by several clinical conditions and through different mechanisms, probably due to different diseases that lead to a susceptible condition with a trigger.<sup>2,8</sup> Some causes may include VT/VF episodes contributing to the recurrence of VT/VF, even with an implantable cardioverter-defibrillator, shock-induced sympathetic activation, acute global ischemia during VT/VF, and shock-induced myocardial damage and dysfunction.<sup>2</sup>

### Specific Diseases

There are a few conditions that can cause ES, some of which are listed below:

#### *Brugada Syndrome*

Brugada syndrome, a genetic sudden cardiac death arrhythmia, is one of the possible causes of ES. It is characterized by ST segment elevation in the right precordial leads, a high incidence of polymorphic VT and VF, and complete or incomplete right bundle-branch block.<sup>2</sup>

This mechanism involves accelerated repolarization caused by abnormalities in the ion channels in certain regions. This results in a lack of an AP dome, prominent action potential duration condensation, transmural voltage gradient, elevation of ST-segment phase 2 reentry, and, ultimately, VT and VF. Another possibility could be the delayed activation of the right ventricular outflow tract due to conduction problems. The syndrome also has some predisposing factors, including hypokalemia, bradycardia, fever, and high vagal tone.<sup>2</sup>

Brugada syndrome's VF can usually be treated with isoprenaline (a beta-agonist) in acute cases and can be prevented with isoprenaline, cilostazol, and denopamine.<sup>2</sup>

#### *Congenital long QT Syndrome*

Congenital long QT syndrome is a genetic condition

that involves prolongation of QT intervals, increased risk of VA, and sudden cardiac death (SCD). It is believed that early depolarization causes muscle contractions due to large spatial gradients in repolarization and repolarization prolongation, leading to block and reentry. Drug therapy is performed using antiadrenergic drugs, although evidence also shows the possibility of progesterone helping to prevent calcium oscillations and early after-depolarization.<sup>2,15</sup>

Additionally, an ICD is also indicated, but can create problems, as there are usually many small episodes of arrhythmia, leading to the overuse of the ICD if the detection time is not sufficiently long. After a shock, sympathetic hyperactivity occurs, increasing the risk of tachyarrhythmia and ES. Thus, reprogramming the ICD is important to avoid extra shocks and may even reduce ES events.

#### *Catecholaminergic Polymorphic Ventricular Tachycardia*

In addition, genetic SCD syndrome is described as a bidirectional or polymorphic VT with an increased sympathetic tone, housed in a structurally normal heart. There are no Electrocardiographic (ECG) manifestations if the patient is in a resting condition.<sup>2</sup>

This is because mutations increase the responsiveness to luminal and/or intracellular calcium levels, leading to delayed afterdepolarization-triggered activity. This condition can interact with an increased sympathetic tone, accentuating the case.<sup>2</sup>

Beta-blocker therapy can be successful in most patients, but some remain with significant arrhythmic events. Other therapies include class 1c antiarrhythmics (propafenone and flecainide) and left cardiac sympathetic denervation. ICD is not indicated, as it can lead to ES.<sup>2</sup>

#### *Hypertrophy and Heart Failure*

Both hypertrophy and heart failure increase the chances of SCD caused by VT and VF, which are risk factors for ES development, while ES is linked to increased mortality. It is presumed that there are two mechanisms related to reentry and focal activity. The reentry theory is supported by several factors, such as fibrosis and ES being suppressed by class III antiarrhythmic azimilide and the effectiveness of catheter ablation. The focal activity theory is considered because three-dimensional mapping often shows VT as a result of focal activity.<sup>2</sup>

The risk of SCD in patients with heart failure can be

reduced using beta-blockers, angiotensin-receptor blockers, lipid-lowering agents, aldosterone receptor inhibitors, and angiotensin-converting enzyme inhibitors. Cardiac resynchronization therapy with biventricular pacing reduces the risk of ES and can reverse ventricular remodeling in Heart Failure (HF).

## Diagnosis and prognosis

### Diagnosis

An integrated assessment of the clinical data and results of complementary examinations is required, as this integration is crucial for the diagnosis of rare diseases. Careful risk stratification and early recognition of unfavorable prognostic signs are of pivotal importance in achieving success.<sup>16</sup> Baseline investigations included electrolytes, Transthoracic Echocardiogram (TTE), and ECG (including QT interval).<sup>17,18</sup> In addition, determining the cause of ES is essential, considering that treatment must target the underlying mechanism.<sup>19,20</sup>

ES can initially be classified based on three gross ECG surface morphologies: monomorphic VT, polymorphic VT, or VF.<sup>18,19,21-23</sup> The mapping was performed using electrode catheters with an interelectrode distance of 5-10 mm and a filter setting of 30-500 Hz. Normal electrographic characteristics include an amplitude of 3 mV or more and a duration of 70 msec or less, while abnormal electrographic characteristics include a duration of 60-70 msec or more, an amplitude of 0.5 mV or less, and the presence of frequency-rich, multiphasic waveforms (fractionated or fragmented electrograms). Split potentials are potentials separated by an isoelectric interval of 30 ms or more.<sup>24,25</sup>

Monomorphic VT accounts for most storm cases, caused by re-entry within a heterogeneous ventricular scar in the context of ischemic and non-ischemic cardiomyopathy. In contrast to monomorphic VT storms related to an identifiable electrophysiological substrate, polymorphic VT and VF storms are most often related to Acute Myocardial Ischemia (AMI), ion channelopathies, or idiopathic VF in patients with structurally normal hearts.<sup>19-23</sup> Twelve-lead electrocardiograph documentation of the triggering of premature ventricular contraction is critical in pre-procedural planning to allow the targeting of the triggering beat with ablation.<sup>19</sup>

VT is defined as tachycardia of 100 bpm or more originating from the ventricle (including the His bundle and the conducting system below the His bundle). Sustained VT is defined as VT persisting for more than

30 seconds or requiring prompt termination prior to that caused by hemodynamic compromise. VT can be classified as monomorphic VT with uniform QRS and polymorphic VT with beat-to-beat variation in QRS morphology. Multiple monomorphic VT is called a pleomorphic VT. Polymorphic VT often leads to VF.<sup>18,19</sup> VT arising from the outflow tract is the most common form of idiopathic VT (OT-VT), which is characterized by VT with a left bundle branch block and inferior axis QRS morphology.<sup>20</sup> However, incessant VT is a condition in which a sustained VT resumes within 5 min after successful ICD therapy and continues for over 12 h.<sup>26,27</sup>

### Prognosis

ES directly affects the prognosis of patients.<sup>19</sup> Patients with ES have a 2-7 times greater risk of sudden death during clinical follow-up, especially considering that ES is recognized as a relevant mortality indicator (35% per year after one ES episode).<sup>9,28,29</sup> In addition, mortality occurs early after ICD implantation.<sup>28,30</sup>

A possible cause for the increased chance of mortality is the deterioration of cardiac function following ICD implantation, although this has not been proven. A prospective cohort of 146 patients corroborates this, as 28% of the patients' CI worsened, leading to death.<sup>31</sup>

Hospitalization is common (80%), but it is not obligatory for the adequate treatment of all ES patients. After each ICD shock, hospitalization rates increase by up to 3.39 times and are associated with a worse quality of life and psychological condition, along with higher operating costs.<sup>31</sup> This is in part caused by recurrent ES episodes, which occur in 50%-81% of patients after the first ES episode.<sup>12</sup>

### Treatment

ES treatment is a multimodal approach with a variety of options, including reprogramming of the ICD, drug therapy, neuraxial modulation, sedation, and CA.<sup>12</sup>

### Emergency Treatment

It is necessary to treat high-and low-risk patients to suppress persistent VT or VF and prevent future ICD shocks. However, risk stratification should be used to evaluate high-risk indicators, such as hemodynamic instability, left ventricular ejection fraction <30%, severe chronic pulmonary obstruction, and moderate or severe renal insufficiency. Patients should be moved to

the intensive care unit if necessary.<sup>12</sup>

### *Ventilation and Sedation*

Sympathetic hyperactivity is linked to ES occurrence; thus, in patients with high-risk non-treatable VA, ventilation and sedation should be considered. Benzodiazepines and short-term analgesics can be a first-line treatment because of their ability to provide both sedation and analgesia without a negative inotropic effect. There are reports that propofol can be used; however, since it can have a negative inotropic effect, caution should be exercised to avoid possible Cardiac Insufficiency (CI).<sup>12,32,33</sup>

### *Drug Therapy*

The basis of drug therapy has long been the use of anti-arrhythmic drugs. A meta-analysis showed a 1.5-fold decrease in ES recurrence in patients who were undergoing drug therapy treatment, although it did not achieve a lower mortality.<sup>34</sup> To select the best drug, it is necessary to comprehend the CI severity, potential toxic effects, and ES etiology. In severe VT, drug therapy has a 7% chance of being pro-arrhythmic.<sup>35</sup>

### *Beta-Blockers*

A higher sympathetic tone is the main factor in the development of VA.<sup>36</sup> In ES with VT, ICD shocks can lead to a higher sympathetic tone, resulting in VA and more shocks. With beta-blocker drug therapy, the tonus can be suppressed, making it the first-line therapy, thereby reducing arrhythmia recurrence by 52%.<sup>37</sup>

In ES cases, there are specific benefits for the use of beta-blockers, as a significant number of ES patients also have chronic cardiac insufficiency and down regulation of  $\beta_1$ -adrenergic receptors. Therefore, propranolol and other non-selective drugs are preferred for treatment. If there is a hemodynamic association, short-action drugs such as esmolol can be used, which is a better choice.<sup>38</sup>

A randomized study with 60 ICD patients who developed ES up to 24 h after admission and were treated with either propranolol or metoprolol and intravenous amiodarone for 48 h demonstrated a lower rate of VA in patients using propranolol (10% propranolol vs. 47% metoprolol), shorter hospitalization time, and time to the end of 38 arrhythmia.

### *Amiodarone*

Amiodarone is used in ES treatment by exerting a

class III effect of blocking potassium channels and prolonging the myocyte refractory period. It can also have a class I effect (by inhibiting sodium channels), class II (blocking L-type calcium channels), and class IV (sympathetic blockage), particularly if administered intravenously.<sup>39</sup>

Amiodarone also succeeds in lowering both the rates and recurrence of VT and, in the long term, it is used in isolation by 40% of the patients. When associating both amiodarone and beta-blockers, ICD recurrent shocks are reduced compared to beta-blockers and Sotalol isolated.<sup>40</sup> It can also lower ES recurrence by four times during the first two years of follow-up, although it has toxic effects including hypothyroidism and hyperthyroidism, pro-arrhythmic effects, pulmonary fibrosis, and hepatic dysfunction. Therefore, continuous use is limited and may be associated with increased mortality.<sup>34,41</sup>

### *Sotalol*

Sotalol has both beta-blocker and class III anti-arrhythmic activity. A randomized study demonstrated that, compared to placebo, sotalol reduced ICD shocks and reduced death by 48%.<sup>42</sup>

### *Class I Antiarrhythmic Drugs*

Class I drugs such as lidocaine, mexiletine, and procainamide are used in ES therapy, but with uncertainty. Their effect is based on the fast inhibition of sodium channels, which makes lidocaine a possible choice for ES caused by IHD.<sup>43</sup> Mexiletine can reduce the frequency of VA and is used in association with amiodarone to reduce ICD shocks, although it can increase mortality<sup>44</sup> and can be toxic to the central nervous system.<sup>12</sup> Procainamide also acts by blocking sodium channels and is more effective and safer than amiodarone.<sup>45</sup>

### *Ablation*

CA destroys a specific area that causes abnormal electric impulse generation or propagation.<sup>4</sup> Most authors indicate the procedure when ES is resistant to drug therapy and as a rescue therapy, considering that both amiodarone and sympathetic blockade can lead to instability or might require more time to reach stability.<sup>46</sup>

### *Types of Mapping*

A wide variety of mapping techniques can be used to

identify the origin of ventricular arrhythmia and select targets for ablation.

### *Activation Mapping*

Activation mapping is used to identify hemodynamically stable focal VT.<sup>5</sup> This method requires an active VT at the same moment as the exam is performed; therefore, it is important to limit VT duration as much as possible to avoid unnecessary risks.<sup>47</sup>

### *Pace Mapping*

Pace mapping is performed by trying to relate the possible pacing site to a re-entry circuit or tachycardia focus, which is performed in the absence of tachycardia.<sup>5</sup>

### *Entrainment Mapping*

Patients with scar-related VT usually suffer from high complexity re-entry circuits and slow conduction regions; therefore, entrainment mapping is preferred instead of activation mapping alone,<sup>5</sup> which is indicated for organized, monomorphic, reentrant VT.<sup>47</sup> Similar to activation mapping, entrainment mapping also requires an active VT at the moment of the exam.<sup>47</sup>

### *Substrate Mapping*

Substrate mapping allows most ablation and mapping to be performed in the sinus rhythm, serving as an option in many patients with structural heart disease and having the possibility of combining it with limited entrainment and activation mapping.<sup>5</sup>

### *Transthoracic Epicardial Mapping and Ablation*

This modality is based on the use of an epidural anesthesia needle to perforate the thorax and perform both mapping and ablation. Irrigated radiofrequency ablation is usually performed because of the higher achievable power delivery due to the lack of circulating blood to cool down the standard radiofrequency catheter. It carries a risk of coronary artery injury.<sup>5</sup>

### *Access Points*

There are two possible routes to access the left ventricle: transaortic (also called retrograde) and transseptal.<sup>46</sup> The transaortic approach is most commonly used, but in some cases (elders, aortic stenosis, artificial aortic valve, vascular disease, or aortic aneurysm), the transseptal approach is preferred.<sup>48</sup>

A study with 21 patients that compared both techniques

showed little success difference, with 83% (transseptal) and 80% (transaortic) acute therapeutic success. Although the effectiveness was similar, there were a few differences between the two approaches.<sup>48</sup>

Patients were divided based on whether it was possible to perform the transaortic route (15 patients) or the transseptal technique (six patients with either aortic stenosis or peripheral vascular insufficiency). One patient was submitted to both, as the aortic procedure failed. In the retrograde group, it was noted that the ventricle volume was overestimated during mapping, probably due to a less stable catheter tip requiring more pressure against the ventricle wall compared to the transseptal group. ICD shocks occurred in one patient in the transseptal group and in three patients in the retrograde group at the months follow-up, although ES was not observed.<sup>48</sup>

Although the transaortic technique is more commonly used, the transseptal approach is precise, safe, and viable.<sup>49</sup>

### *Types of Approaches to Ablation*

Depending on the underlying disease, different types of ablation can be performed. Ischemic Cardiomyopathy (ICM) usually requires endocardial ablation, while non-ischemic cardiomyopathy is treated using an epicardial approach.<sup>46</sup>

The optimal approach for ablation depends on the arrhythmia mechanism, as it can be either focal or macro re-entrant and can be caused by different substrates. These include non-ischemic and ischemic cardiomyopathy, prior cardiac surgery, sarcoidosis, Chagas disease etc. Usually, the possible location is first determined by ECG, then the underlying disease should be diagnosed when possible, and each condition follows its own diagnostic path.<sup>50</sup>

Additionally, in a study on 218 patients (mean age, 59 years) and 1836 VT ablations, epicardial ablation (or a combined endo-epicardial approach) was indicated if the VT was likely epicardial due to the ECG result of spontaneous VT or underlying disease, there was a lack of significant areas of low voltage at sinus rhythm on endocardial mapping or an electrogram, a “pseudofocal” pattern of endocardial activation, or an intracardiac thrombus or valve prosthesis (mitral or aortic) was present. Epicardial ablation was the first choice in 78 patients (35.8%), followed by failure of endocardial ablation in the remaining 140 patients (64.2%).<sup>51</sup>

### Radiofrequency

Radiofrequency was first introduced in 1978 and is now one of the most commonly used energy sources. Alternating current with continuous sinusoidal unmodulated waveform (300-1000 kHz) is more frequent, allowing the generation of myocardial lesions that are well circumscribed. This is done by heating the local, and the ablated region depends on the duration and temperature.<sup>5</sup>

### Irrigated Radiofrequency Ablation

Irrigated radiofrequency ablation is based on cooling the catheter with either circulating fluids inside the electrode (closed loop) or by flushing saline through pores in the electrode (open irrigation). This permits higher energy delivery, although large amounts of administered saline can lead to either respiratory or hemodynamic compromise.<sup>5</sup>

### Pulsed Radiofrequency Ablation

This is done by pulsing energy for a few milliseconds, allowing greater energy levels at the same temperature and the possibility of larger lesion depths.<sup>5</sup>

### Alcohol Ablation

Alcohol is cytotoxic, leading to irreversible tissue damage due to necrosis and coronary vessel occlusion. First, the coronary artery that supplies the target area was identified, and then ethanol was injected. This technique is moderately successful and has some risks, including lesions in other myocardium areas.<sup>5</sup>

### Cryoablation

It can produce lesions smaller than radiofrequency, possibly causing less damage to adjacent structures.<sup>5</sup>

### Success or Fail

The ablation results were classified into three categories. Absolute success was considered when there was no induction of any clinical VT when the procedure was finalized (programmed pacing protocol), whereas partial success was defined by the absence of clinical VT. Finally, failure is the possibility of clinical VT induction.<sup>46</sup>

### Rates of Success

In a study of 40 patients and 84 total VTs induced ( $2.1 \pm 1$  procedure per patient), VTs were successfully

ablated using remote magnetic navigation in 95% of patients, and ES was eliminated during the observation period (48 h). The rates did not vary between patients treated with or without AMD, and no major complications occurred during observations. In long term ( $17.4 \pm 16.9$  months), 47.5% had no recurrence of sustained VT and these patients did not receive ICD therapy. The ICD usage percentage before ablation was 69%, whereas it was only 30% after ablation ( $P < 0.01$ ), and the number of ICD shocks diminished by 56% compared to before the ablation ( $P < 0.05$ ).<sup>49</sup>

A systematic review of 11 articles, also about remote magnetic navigation, found that acute success for ICM varied from 36% to 80%, while non-ischemic success was between 50% and 100%, and the lowest and highest structural normal heartsuccess rates were 86% and 100%, respectively. In long term, recurrence for ICM was 0%-30%, for non-ischemic was 14%-50% (although these data were from 16 patients in only two studies), and for normal structure was 8%-17%.<sup>52-54</sup>

### Complications and Predictors

Complications are divided into two categories: major and minor. Major complications lead to long-term disability, requiring hospitalization or increased hospitalization. Minor complications are more easily dealt with, including those related to complaints at puncture sites and pericarditis, although some authors incorporate transient total atrioventricular block, transient acute heart failure, and left bundle branch block as minor complications. Cardiac tamponade is the most feared complication, although it is infrequent.<sup>46</sup>

In a study of 548 patients who underwent 722 ablation procedures (29% indicated for ES), 45 (6.2%) procedures resulted in major complications. The most frequent complications were vascular injury (58% of total complications), and perforation (tamponade and hemopericardium) were present in 0.4% of cases, while conduction system damage was present in 1% of cases. Finally, the incidence of pericarditis was 0.13%. Different diseases and clinical conditions require different approaches.<sup>55</sup>

The same study found a few predictors for complications that were statistically relevant for CA in general and not only indicated ES. These included high-risk operators ( $P = 0.0001$ ), age  $> 70$  years ( $P = 0.003$ ), arterial access ( $P = 0.008$ ), non-elective procedure ( $P = 0.01$ ), heart failure ( $P = 0.0006$ ), ICD ( $P = 0.04$ ), and serum

creatinine >115  $\mu\text{mol/L}$  ( $P = 0.00007$ ), among others. It is worth mentioning that ES was not statistically significant ( $P = 0.05$ ).<sup>55,56</sup>

### **Alternative Treatments**

In cases of cardiac ES refractory to the treatments, some alternative methods have been proposed, such as variations of radiofrequency ablation methods including sequential, simultaneous, and bipolar ablation, which will expand the lesion size in order and thus reach deep intramural VT. Variations in radiofrequency ablation methods such as sequential, simultaneous, and bipolar ablation will expand the lesion size and thus reach deep intramural VTs. The new technique of needle ablation involves plunging a small 27-gauge electrode up to 1 cm into the myocardium for intramural ablation. Surgical cryoablation has been reported to be effective in patients with non-ischemic cardiomyopathy and ventricular tachycardia refractory to standard therapy. Trans-coronary ethanol ablation is an alternative therapy for refractory VT in patients with structural heart disease. Transverse ethanol ablation has also been reported as an alternative therapy for intramural VT from the left ventricle summit and is best performed by experts in this technique.<sup>12</sup>

### **Neuraxial Modulation**

The increase in sympathetic tonus is an important factor at the beginning of ES and during potentialization. Therefore, modulating elements inside the cardiac neuro-axis can be used in the treatment of VA, especially ES.<sup>36,57-63</sup>

Neuraxial modulation can be achieved by Thoracic Epidural Anesthesia (TEA), Renal Denervation (RDN), Stellate Ganglion Blockade (SGB), and Cardiac Sympathetic Denervation (CSD).<sup>59,61</sup>

TEA is based on percutaneous administration of 0.25% bupivacaine (1 ml), followed by 2 ml/h infusion in the thoracic epidural space. It is a short-term treatment that works as a bridge until a definitive approach is reached.

SGB is performed by injecting a local anesthetic on both sides or the left side of the ganglia, leading to the blocking of both afferent and efferent neurons, thereby diminishing the sympathetic flux to the heart. Similar to TEA, this is the stop gap.

CSD is a type of permanent neuroaxis blockade that affects efferent and afferent heart innervation. This resects the stellate ganglion's low half or third and the

2nd, 3rd, and 4th paravertebral ganglia.<sup>60-63</sup>

Lastly, the RDN consists of ablating the neural plexus around the renal artery. This decreases the sympathetic flux to the heart by reducing norepinephrine levels and inhibiting the renin-aldosterone system.

### **Discussion and Conclusion**

An electrical storm is an arrhythmic emergency and life-threatening condition characterized by severe electrical instability. Its mechanism has not been fully elucidated, as it can be triggered by several clinical conditions and through different mechanisms. Early recognition and appropriate management are of paramount importance and greatly impact the patient's survival, and should be performed in an emergency setting by a team that offers a structured and multidisciplinary approach. Determining the cause of ES which is essential, considering that treatment must target the underlying mechanism. A diagnostic approach based on the type of ventricular arrhythmia and the presence or absence of structural heart disease facilitates therapeutic management.

These patients require treatment, including reprogramming of the implantable cardioverter defibrillator, drug therapy, neuraxial modulation, ventilation, sedation, and catheter ablation. A wide variety of mapping techniques have been developed over the years; these techniques can be used in an attempt to identify the origin of a ventricular arrhythmia and select targets for ablation that depend on the underlying disease; moreover, they can be performed using different approaches. Furthermore, in selected cases of cardiac ES refractory to or unsuitable for catheter ablation, some alternative methods have been proposed, such as surgical cryoablation or transcoronary ethanol ablation. Although there is clinical improvement in patients due to advances in non-pharmacological treatments, the ideal pharmacological treatment remains essential for a successful management of ES.

### **Acknowledgement**

The authors are grateful to the School of Medical, Pharmaceutical and Biomedical Sciences of Pontifical Catholic University of Goiás (PUC GO) for supporting this work.

### **Conflict of Interest**

The authors declare that they have no conflicts interest.

## References

- Conti S, Pala S, Biagioli V, Del Giorno G, Zucchetti M, Russo E, et al. Electrical storm: A clinical and electrophysiological overview. *World J Cardiol.* 2015;7(9):555-61. doi:10.4330/wjc.v7.i9.555
- Tsuji Y, Heijman J, Nattel S, Dobrev D. Electrical storm: recent pathophysiological insights and therapeutic consequences. *Basic Res Cardiol.* 2013;108(2):336. doi:10.1007/s00395-013-0336-2
- Israel CW, Manegold JC. Elektrischer Sturm. *Herzschrittmachtherapie + Elektrophysiologie.* 2014;25(2):59-65.
- Harrison's principles of internal medicine. 19th ed. New York: The McGraw-Hill Companies, Inc.; 2015.
- Natale A, Raviele A, AL-AHMAD AM, Alfieri O, Aliot E, Almendral J, et al. Venice Chart International Consensus document on ventricular tachycardia/ventricular fibrillation ablation. *J Cardiovasc Electrophysiol.* 2010;21(3):339-79. doi:10.1111/j.1540-8167.2009.01686.x
- Kuck KH, Schaumann A, Eckardt L, Willems S, Ventura R, Delacretaz E, et al. Catheter ablation of stable ventricular tachycardia before defibrillator implantation in patients with coronary heart disease (VTACH): a multicentre randomised controlled trial. *Lancet.* 2010;375(9708):31-40. doi:10.1016/S0140-6736(09)61755-4
- Calkins H, Epstein A, Packer D, Arria AM, Hummel J, Gilligan DM, et al. Catheter ablation of ventricular tachycardia in patients with structural heart disease using cooled radiofrequency energy: results of a prospective multicenter study. *J Am Coll Cardiol.* 2000;35(7):1905-14. doi:10.1016/s0735-1097(00)00615-x
- Hohnloser SH, Al-Khalidi HR, Pratt CM, Brum JM, Tatla DS, Tchou P, et al. Electrical storm in patients with an implantable defibrillator: incidence, features, and preventive therapy: insights from a randomized trial. *Eur Heart J.* 2006;27(24):3027-32. doi:10.1093/eurheartj/ehl276
- Noda T, Kurita T, Nitta T, Chiba Y, Furushima H, Matsumoto N, et al. Significant impact of electrical storm on mortality in patients with structural heart disease and an implantable cardiac defibrillator. *Int J Cardiol.* 2018;255:85-91. doi:10.1016/j.ijcard.2017.11.077
- Kim SG, Ling J, Fisher JD, Wang G, Rameneni A, Roth JA, et al. Comparison and frequency of ventricular arrhythmias after defibrillator implantation by thoracotomy versus nonthoracotomy approaches. *Am J Cardiol.* 1994;74(12):1245-8. doi:10.1016/0002-9149(94)90556-8
- Credner SC, Kligenheben T, Mauss O, Sticherling C, Hohnloser SH. Electrical storm in patients with transvenous implantable cardioverter-defibrillators: incidence, management and prognostic implications. *J Am Coll Cardiol.* 1998;32(7):1909-15. doi:10.1016/s0735-1097(98)00495-1
- Geraghty L, Santangeli P, Tedrow UB, Shivkumar K, Kumar S. Contemporary management of electrical storm. *Heart Lung Circ.* 2019;28(1):123-33. doi:10.1016/j.hlc.2018.10.005
- Pereira FT, Rocha EA, Monteiro MD, Lima ND, Rodrigues Sobrinho CR, Pires Neto RD. Clinical course after cardioverter-defibrillator implantation: Chagasic versus ischemic patients. *Arq Bras Cardiol.* 2016;107:99-100. doi:10.5935/abc.20160101
- Huang DT, Traub D. Recurrent ventricular arrhythmia storms in the age of implantable cardioverter defibrillator therapy: a comprehensive review. *Prog Cardiovasc Dis.* 2008;51(3):229-36. doi:10.1016/j.pcad.2008.07.003
- Moss AJ. Sex hormones and ventricular tachyarrhythmias in LQTS: new insights regarding antiarrhythmic therapy. *Heart Rhythm.* 2012;9(5):833-4. doi:10.1016/j.hrthm.2012.01.014
- Muser D, Liang JJ, Santangeli P. Electrical storm in patients with implantable cardioverter-defibrillators: a practical overview. *J Innov Card Rhythm Manag.* 2017;8(10):2853-61. doi:10.19102/icrm.2017.081002
- Godinho AR, Frutuoso C, Vasconcelos M, Dias P, Garcia R, Pinho T, et al. Arrhythmic storm: Short-coupled variant torsade de pointes. *Rev Port Cardiol.* 2016;35(5):307.e1-e5. doi:10.1016/j.repc.2016.01.015
- Maruyama M. Management of electrical storm: The mechanism matters. *J Arrhythm.* 2014;30(4):242-9. doi:10.1016/j.joa.2014.03.012
- Haegeli LM, Della Bella P, Brunckhorst CB. Management of a patient with electrical storm: role of epicardial catheter ablation. *Circulation.* 2016;133(7):672-6. doi:10.1161/CIRCULATIONAHA.115.016336
- Eifling M, Razavi M, Massumi A. The evaluation and management of electrical storm. *Texas Heart Inst J.* 2011;38(2):111-21.
- Murata H, Miyauchi Y, Hayashi M, Iwasaki YK, Yodogawa K, Ueno A, et al. Clinical and electrocardiographic characteristics of electrical storms due to monomorphic ventricular tachycardia refractory to intravenous amiodarone. *Circ J.* 2015;79(10):2130-7. doi:10.1253/circj.CJ-15-0213
- Viskin S, Belhassen B. Polymorphic ventricular tachyarrhythmias in the absence of organic heart disease: classification, differential diagnosis, and implications for therapy. *Prog Cardiovasc Dis.* 1998;41(1):17-34. doi:10.1016/S0033-0620(98)80020-0
- Aliot EM, Stevenson WG, Almendral-Garrote JM, Bogun F, Calkins CH, Delacretaz E, et al. EHRA/HRS expert consensus on catheter ablation of ventricular arrhythmias: developed in a partnership with the European Heart Rhythm Association (EHRA), a registered branch of the European Society of Cardiology (ESC), and the Heart Rhythm Society (HRS); in collaboration with the American College of Cardiology (ACC) and the American Heart Association (AHA). *Europace.* 2009;11(6):771-817. doi:10.1093/europace/eup098
- Cassidy DM, Vassallo JA, Marchlinski FE, Buxton AE, Untereker WJ, Josephson ME. Endocardial mapping in humans in sinus rhythm with normal left ventricles: activation patterns and characteristics of electrograms. *Circulation.* 1984;70(1):37-42. doi:10.1161/01.CIR.70.1.37
- Stevenson WG, Wilber DJ, Natale A, Jackman WM, Marchlinski FE, Talbert T, et al. Irrigated radiofrequency catheter ablation guided by electroanatomic mapping for recurrent ventricular tachycardia after myocardial infarction: the multicenter thermocool ventricular tachycardia ablation trial. *Circulation.* 2008;118(25):2773-82. doi:10.1161/CIRCULATIONAHA.108.788604
- Hendriks AA, Szili-Torok T. Editor's Choice-The treatment of electrical storm: an educational review. *Eur Heart J Acute Cardiovasc Care.* 2018;7(5):478-83. doi:10.1177/2048872618781358
- Wu CI, Chang SL, Lin CY, Vicera JJ, Lin YJ, Lo LW, et al. Clinical significance of J wave in prediction of ventricular arrhythmia in patients with acute myocardial infarction. *J Cardiol.* 2019;73(5):351-7. doi:10.1016/j.jjcc.2018.11.005
- Sesselberg HW, Moss AJ, McNitt S, Zareba W, Daubert JP, Andrews ML, et al. Ventricular arrhythmia storms in postinfarction patients with implantable defibrillators for primary prevention indications: a MADIT-II substudy. *Heart Rhythm.* 2007;4(11):1395-402. doi:10.1016/j.hrthm.2007.07.013
- Izquierdo M, Ruiz-Granell R, Ferrero A, Martínez A, Sanchez-Gomez J, Bonanad C, et al. Ablation or conservative management of electrical storm due to monomorphic ventricular tachycardia: differences in outcome. *Europace.* 2012;14(12):1734-9. doi:10.1093/europace/eus186
- Guerra F, Flori M, Bonelli P, Patani F, Capucci A. Electrical storm and heart failure worsening in implantable cardiac defibrillator patients. *Ep Europace.* 2014;17(2):247-54. doi:10.1093/europace/euu298
- Mandel JE, Hutchinson MD, Marchlinski FE. Remifentanyl-midazolam sedation provides hemodynamic stability and comfort during epicardial ablation of ventricular tachycardia. *J Cardiovasc Electrophysiol.* 2011;22(4):464-6. doi:10.1111/j.1540-8167.2010.01889.x
- Mulpuru SK, Patel DV, Wilbur SL, Vasavada BC, Furqan T. Electrical storm and termination with propofol therapy: a case report. *Int J Cardiol.* 2008;128(1):e6-8. doi:10.1016/j.ijcard.2007.05.052
- Santangeli P, Muser D, Maeda S, Filtz A, Zado ES, Frankel DS, et al. Comparative effectiveness of antiarrhythmic drugs and catheter ablation for the prevention of recurrent ventricular tachycardia in patients with implantable cardioverter-defibrillators: a systematic review and meta-analysis of randomized controlled trials. *Heart Rhythm.* 2016;13(7):1552-9. doi:10.1016/j.hrthm.2016.03.004
- Stanton MS, Prystowsky EN, Fineberg NS, Miles WM, Zipes DP, Heger JJ. Arrhythmogenic effects of antiarrhythmic drugs: a

- study of 506 patients treated for ventricular tachycardia or fibrillation. *J Am Coll Cardiol.* 1989;14(1):209-15.
35. Vaseghi M, Gima J, Kanaan C, Ajijola OA, Marmureanu A, Mahajan A, et al. Cardiac sympathetic denervation in patients with refractory ventricular arrhythmias or electrical storm: intermediate and long-term follow-up. *Heart Rhythm.* 2014;11(3):360-6. doi:10.1016/j.hrthm.2013.11.028
  36. Moss AJ, Zareba W, Hall WJ, Klein H, Wilber DJ, Cannom DS, et al. Prophylactic implantation of a defibrillator in patients with myocardial infarction and reduced ejection fraction. *New Engl J Med.* 2002;346(12):877-83. doi:10.1056/NEJMoa013474
  37. Chatzidou S, Kontogiannis C, Tsilimigras DI, Georgiopoulos G, Kosmopoulos M, Papadopoulou E, et al. Propranolol versus metoprolol for treatment of electrical storm in patients with implantable cardioverter-defibrillator. *J Am Coll Cardiol.* 2018;71(17):1897-906. doi:10.1016/j.jacc.2018.02.056
  38. Amiodarone Trials Meta-Analysis Investigators. Effect of prophylactic amiodarone on mortality after acute myocardial infarction and in congestive heart failure: meta-analysis of individual data from 6500 patients in randomised trials. *Lancet.* 1997;350(9089):1417-24. doi:10.1016/S0140-6736(97)05281-1
  39. Connolly SJ, Dorian P, Roberts RS, Gent M, Bailin S, Fain ES, et al. Comparison of  $\beta$ -blockers, amiodarone plus  $\beta$ -blockers, or sotalol for prevention of shocks from implantable cardioverter defibrillators: the OPTIC study: a randomized trial. *JAMA.* 2006;295(2):165-71. doi:10.1001/jama.295.2.165
  40. Huang SK, de Guzman WL, Chenarides JG, Okike NO, Vander Salm TJ. Effects of long-term amiodarone therapy on the defibrillation threshold and the rate of shocks of the implantable cardioverter-defibrillator. *Am Heart J.* 1991;122(3):720-7. doi:10.1016/0002-8703(91)90517-L
  41. Pacifico A, Hohnloser SH, Williams JH, Tao B, Saksena S, Henry PD, et al. Prevention of implantable-defibrillator shocks by treatment with sotalol. *New Engl J Med.* 1999;340(24):1855-62. doi:10.1056/NEJM199906173402402
  42. Nasir Jr N, Taylor A, Doyle TK, Pacifico A. Evaluation of intravenous lidocaine for the termination of sustained monomorphic ventricular tachycardia in patients with coronary artery disease with or without healed myocardial infarction. *Am J Cardiol.* 1994;74(12):1183-6. doi:10.1016/0002-9149(94)90544-4
  43. Gao D, Van Herendael H, Alshengeiti L, Dorian P, Mangat I, Korley V, et al. Mexiletine as an adjunctive therapy to amiodarone reduces the frequency of ventricular tachyarrhythmia events in patients with an implantable defibrillator. *J Cardiovasc Pharmacol.* 2013;62(2):199-204. doi:10.1097/FJC.0b013e31829651fe
  44. Ortiz M, Martin A, Arribas F, Coll-Vinent B, Del Arco C, Peinado R, et al. Randomized comparison of intravenous procainamide vs. intravenous amiodarone for the acute treatment of tolerated wide QRS tachycardia: the PROCAMIO study. *Eur Heart J.* 2017;38(17):1329-35. doi:10.1093/eurheartj/ehw230
  45. Leal RT, Monteiro GC, Junior AD. Catheter ablation in the treatment of electrical storm: Integrative review. *Indian Pacing Electrophysiol J.* 2017;17(5):140-5. doi:10.1016/j.ipej.2017.07.012
  46. Kumar S, Tedrow UB, Stevenson WG. Entrainment mapping. *Card Electrophysiol Clin.* 2017;9(1):55-69. doi:10.1016/j.ccep.2016.10.004
  47. Pluta S, Lenarczyk R, Pruszkowska-Skrzep P, Kowalski O, Sokal A, Sredniawa B, et al. Transseptal versus transaortic approach for radiofrequency ablation in patients with cardioverter-defibrillator and electrical storm. *J Interv Card Electrophysiol.* 2010;28(1):45-50. doi:10.1007/s10840-009-9464-x
  48. Jin Q, Jacobsen PK, Pehrson S, Chen X. Acute and long term outcomes of catheter ablation using remote magnetic navigation for the treatment of electrical storm in patients with severe ischemic heart failure. *Int J Cardiol.* 2015;183:11-6. doi:10.1016/j.ijcard.2015.01.066
  49. Yamada T, Kay GN. Optimal ablation strategies for different types of ventricular tachycardias. *Nat Rev Cardiol.* 2012;9(9):512-25. doi:10.1038/nrcardio.2012.74
  50. Della Bella P, Brugada J, Zeppenfeld K, Merino J, Neuzil P, Maury P, Maccabelli G, Vergara P, Baratto F, Berrueto A, Wijngaarden AP. Epicardial ablation for ventricular tachycardia: a European multicenter study. *Circ Arrhythm Electrophysiol.* 2011;4(5):653-9. doi:10.1161/CIRCEP.111.962217
  51. Akca F, Unsesveren I, Jordaens L, Szili-Torok T. Safety and efficacy of the remote magnetic navigation for ablation of ventricular tachycardias—a systematic review. *J Interv Card Electrophysiol.* 2012;34(1):65-71. doi:10.1007/s10840-011-9645-2
  52. Deneke T, Shin DI, Lawo T, Bosche L, Balta O, Anders H, et al. Catheter ablation of electrical storm in a collaborative hospital network. *Am J Cardiol.* 2011;108(2):233-9. doi:10.1016/j.amjcard.2011.03.030
  53. Dello Russo A, Casella M, Pelargonio G, Santangeli P, Bartoletti S, Bencardino G, et al. Feasibility and safety of catheter ablation of electrical storm in ischemic dilated cardiomyopathy. *J Cardiovasc Med.* 2016;17(6):425-32. doi:10.2459/JCM.0000000000000259
  54. Peichl P, Wichterle D, Pavlu L, Cihak R, Aldhoon B, Kautzner J. Complications of catheter ablation of ventricular tachycardia: a single-center experience. *Circ Arrhythm Electrophysiol.* 2014;7(4):684-90. doi:10.1161/CIRCEP.114.001530
  55. Sohrabi B, Mokhtari Nouri J, Moradian ST, Ghiasi SM. Clinical Experiences of the Adaptive Support Ventilation Mode in Cardiac Surgery Patients. *Int J Med Rev.* 2015;2(1):191-5.
  56. Bohnen M, Stevenson WG, Tedrow UB, Michaud GF, John RM, Epstein LM, et al. Incidence and predictors of major complications from contemporary catheter ablation to treat cardiac arrhythmias. *Heart Rhythm.* 2011;8(11):1661-6. doi:10.1016/j.hrthm.2011.05.017
  57. Meng L, Tseng CH, Shivkumar K, Ajijola O. Efficacy of stellate ganglion blockade in managing electrical storm: a systematic review. *JACC Clin Electrophysiol.* 2017;3(9):942-9. doi:10.1016/j.jacep.2017.06.006
  58. Barron HV, Lesh MD. Autonomic nervous system and sudden cardiac death. *J Am Coll Cardiol.* 1996;27(5):1053-60. doi:10.1016/0735-1097(95)00615-X
  59. Bradfield JS, Ajijola OA, Vaseghi M, Shivkumar K. Mechanisms and management of refractory ventricular arrhythmias in the age of autonomic modulation. *Heart Rhythm.* 2018;15(8):1252-60. doi:10.1016/j.hrthm.2018.02.015
  60. Bourke T, Vaseghi M, Michowitz Y, Sankhla V, Shah M, Swapna N, et al. Neuraxial modulation for refractory ventricular arrhythmias: value of thoracic epidural anesthesia and surgical left cardiac sympathetic denervation. *Circulation.* 2010;121(21):2255-62. doi:10.1161/CIRCULATIONAHA.109.929703
  61. Kumar S, Barbhaiya CR, Sobieszczyk P, Eisenhauer AC, Couper GS, Nagashima K, et al. Role of alternative interventional procedures when endo-and epicardial catheter ablation attempts for ventricular arrhythmias fail. *Circ Arrhythm Electrophysiol.* 2015;8(3):606-15. doi:10.1161/CIRCEP.114.002522
  62. Hussain SM, Harky A. Complications of coronary artery bypass grafting. *Int J Med Rev.* 2019;6(1):1-5. doi:10.29252/IJMR-060101
  63. Prabhu MA, Prasad SB, Abhilash SP, Thajudeen A, Namboodiri N. Left sympathetic cardiac denervation in managing electrical storm: acute outcome and long term follow up. *J Interv Card Electrophysiol.* 2016;47(3):285-92. doi:10.1007/s10840-016-0153-2
  64. Kambile FK, Trushin SN, Mikheev AV, Andryushin AS. Postoperative Cardiovascular Morbidity and Management after Lung Surgery: A Retrospective Study and Literature Review. *Int J Med Rev* 2021;8(1):3-11. doi:10.30491/IJMR.2020.234460.1107