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Review

Management of electrical storm: The mechanism matters

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ABSTRACT

An electrical storm is a life-threatening syndrome that is characterized by clustering of recurrent episodes of ventricular tachycardia (VT) or ventricular fibrillation (VF) within a relatively short period of time. Electrical storms occur in a wide variety of conditions, and successful treatment depends on a correct understanding of the mechanism underlying the recurrent arrhythmias. Management of electrical storms is challenging, but classifying patients according to the type of recurrent arrhythmia (monomorphic VT or polymorphic VT/VF) and the presence or absence of structural heart disease would aid differential diagnosis and allow for more specific therapies.

Contents

1. Introduction

The term “electrical storm” describes a state of electrical instability of the heart characterized by clustering of recurrent episodes of ventricular tachycardia (VT) or ventricular fibrillation (VF) in a short period of time. In recent years, implantable cardioverter defibrillators (ICDs) have significantly improved the survival of patients with VT/VF. However, electrical storms remain associated with high mortality and morbidity, and have a negative impact on long-term outcomes [1]. Although there is no consensus regarding the definition of an electrical storm, the generally accepted definition in clinical practice and recent literature is the occurrence of ≥ 2 separate VT/VF episodes or ≥ 3 appropriate ICD therapies for VT/VF in a 24-h period [2]. In more than half of the patients with an electrical storm, the intervals between the VT/VF episodes are < 1 h, with the shortest interval of < 1 min [3]. A sustained VT/VF that resumes immediately after (≥ 1 sinus cycle) successful defibrillation/cardioversion is regarded as a severe form of electrical storm. This may masquerade as shock-refractory VT/VF, since a brief sinus period followed by immediate VT/VF recurrence can be concealed by post-shock electrocardiographic (ECG) saturation [4]. Therefore, a severe form of electrical storm might also be responsible for the events that occur in some patients with shock-refractory VT/VF, which are commonly seen during cardiopulmonary resuscitation. It is imperative to improve the treatment strategy for electrical storms, because the incidence

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Table 1
Causes of electrical storms.

Structural heart diseases
Ischemic heart diseases
Acute or recent myocardial infarction
Prior myocardial infarction
Non-ischemic cardiomyopathy
Dilated cardiomyopathy
Hypertrophic cardiomyopathy
Arrhythmogenic right ventricular dysplasia/cardiomyopathy
Valvular heart diseases
Corrected congenital heart diseases
Myocarditis
Cardiac sarcoidosis
Chagas disease
Metastatic cardiac tumor
Abnormal electrical substrate (structurally normal hearts)
Primary causes
Idiopathic
Brugada syndrome
Early repolarization syndrome
Long QT syndrome
Short QT syndrome
Catecholaminergic polymorphic ventricular tachycardia
Secondary causes
Electrolyte abnormalities
Toxic/drug related
Endocrinologic
Perioperative
Iatrogenic (T wave pacing)

of electrical storms is not low (10–28% in patients with an ICD implanted for secondary prevention and 4% for primary prevention) [1].

This review discusses the practical and efficacious management of electrical storms. Here, I emphasize that the treatment outcome greatly depends on the understanding of the pathophysiology of electrical storms. An approach tailored to the underlying mechanism seems essential for successful management.

2. Principles of management

Patients who present with an electrical storm often have structural heart disease; however, functional electrical abnormalities also provoke electrical storms in patients with structurally normal hearts. The underlying conditions that cause electrical storms are listed in Table 1.

Since an electrical storm is a medical emergency, it is generally managed in line with a treatment algorithm, typically the Advanced Cardiovascular Life Support (ACLS) protocol [5], regardless of the etiology of the electrical storm. Such a pre-specified algorithmic approach is highly effective in critical patient care, which requires a prompt response to life-threatening conditions. Nevertheless, I propose that treatment should be as specific to the underlying mechanism as possible, because electrical storms of some etiologies actually require a completely opposite treatment. Specifically, sympathetic blockade is effective in controlling electrical storms in patients with the majority of structural heart diseases [6–8], congenital long QT syndrome (LQTS) [9], and catecholaminergic polymorphic VT (CPVT) [10], whereas sympathetic stimulation with isoproterenol is useful for inhibiting electrical storms caused by Brugada syndrome [11], early repolarization syndrome [12], and short QT syndrome (SQTS) [13]. Alternatively, antiarrhythmic drugs commonly used for the treatment of electrical storms [5] can aggravate the situation in LQTS by further lengthening the QT interval [14]. Therefore, an empirical approach to treat recurrent VT/VF is potentially harmful in certain cases, and a mechanism-directed therapy would be preferable. Although it is difficult to perform a complete diagnostic evaluation during an electrical storm, standard workups such as transthoracic echocardiography and 12-lead ECG can help one understand whether patients have structural heart disease, and/or some specific ECG findings including ischemic ST-T changes, abnormal QT interval, Brugada-type ECG, and others. Myocardial ischemia, worsening heart failure, and electrolyte disturbances are a common trigger of electrical storms [15]. If such an inciting factor is found, emergent revascularization, correcting the electrolyte abnormality or treatment for heart failure must be performed.

The type of ventricular arrhythmia (monomorphic VT or polymorphic VT/VF) responsible for the electrical storm provides an important diagnostic clue to its pathophysiology (Fig. 1). In this review, we address the diagnosis and specific treatments according to the type of ventricular arrhythmia and the presence or absence of structural heart disease.

3. Monomorphic VT storms in structurally normal hearts

The majority of patients with monomorphic VT storm have structural heart disease, but monomorphic VT storms can occur in structurally normal hearts on rare occasions [16,17] (Fig. 2).

Monomorphic VT occurring in structurally normal hearts is referred to as idiopathic VT. The characteristics of idiopathic VT depend on the origin of the VT. VT arising from the outflow tract is the most common form of idiopathic VT (OT-VT), which is characterized by VT with left bundle branch block (LBBB) and inferior axis QRS morphology. The typical mechanism of OT-VT involves triggered activity due to cyclic adenosine monophosphate (AMP)-mediated delayed afterdepolarizations (DADs) [18]. Beta-adrenergic stimulation increases the intracellular cyclic AMP and intracellular Ca^{2+} levels, resulting in spontaneous Ca^{2+} release from the sarcoplasmic reticulum (SR), DADs, and triggered activity. Therefore, OT-VT can be suppressed typically by beta-blockers that lower the stimulated levels of cyclic AMP (and thus decrease intracellular Ca^{2+}) or non-dihydropyridine Ca^{2+} channel blockers (verapamil or diltiazem) that directly reduce intracellular Ca^{2+} by inhibiting the inward L-type Ca^{2+} current. As a second-line therapy, class IC or III antiarrhythmic agents are also effective for suppressing OT-VT [19]. Radiofrequency catheter ablation (RFCA) is a safe and reliable technique for the treatment of OT-VT, unless it has an epicardial origin that may require less safe approaches through the coronary venous system, a subxyphoid approach for RFCA, or surgical ablation (an epicardial origin is suggested by delayed initial precordial QRS activation as quantified by a maximum deflection index ≥ 0.55 [20], Q wave amplitude in aVL to aVR > 1.4, or S wave amplitude in V1 ≥ 1.2 mV [21]). If available, RFCA can be the first choice of treatment for OT-VT not only when OT-VT is refractory to medical therapy, because RFCA can be a curative treatment for OT-VT. Activation mapping is useful when seeking the VT origin during an electrical storm. The earliest activation at successful ablation sites typically precedes the surface QRS onset by 20–40 ms, with unipolar electrograms exhibiting a QS pattern with a rapid initial downstroke [22].

Fascicular VT, also known as idiopathic left VT, is the second leading cause of idiopathic VT. The mechanism of fascicular VT is supposed to be macro-reentry involving the Purkinje fiber network, which connects to the left fascicle [23,24]. Fascicular VT is sub-classified based on the ECG morphology (a right bundle branch block [RBBB] pattern and superior or inferior QRS axis) and corresponding fascicle coupled to the reentrant circuit: left posterior fascicular VT, left anterior fascicular VT, and left upper septal VT [23]. Left posterior fascicle VT is the most common manifestation. Fascicular VT has a characteristic ECG with a relatively narrow QRS width that reflects the rapid spread of